Pamela Shaw

List of Publications by Citations

406 papers	25,389 citations	87 h-index	143 g-index
450 ext. papers	29,950 ext. citations	7.3 avg, IF	6.81 L-index

#	Paper	IF	Citations
406	Frequency of the C9orf72 hexanucleotide repeat expansion in patients with amyotrophic lateral sclerosis and frontotemporal dementia: a cross-sectional study. *Lancet Neurology, The*, **2012**, 11, 323-30	24.1	830
405	Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurology, The*, **2006**, 5, 140-7	24.1	704
404	Pathological TDP-43 distinguishes sporadic amyotrophic lateral sclerosis from amyotrophic lateral sclerosis with SOD1 mutations. *Annals of Neurology*, **2007**, 61, 427-34	9.4	698
403	Adaptation to culture of human embryonic stem cells and oncogenesis in vivo. *Nature Biotechnology*, **2007**, 25, 207-15	44.5	520
402	Amyotrophic lateral sclerosis. *Nature Reviews Disease Primers*, **2017**, 3, 17071	51.1	459
401	Molecular pathways of motor neuron injury in amyotrophic lateral sclerosis. *Nature Reviews Neurology*, **2011**, 7, 616-30	15	428
400	White matter lesions in an unselected cohort of the elderly: molecular pathology suggests origin from chronic hypoperfusion injury. *Stroke*, **2006**, 37, 1391-8	6.7	416
399	Oxidative stress in ALS: key role in motor neuron injury and therapeutic target. *Free Radical Biology and Medicine*, **2010**, 48, 629-41	7.8	415
398	Controversies and priorities in amyotrophic lateral sclerosis. *Lancet Neurology, The*, **2013**, 12, 310-22	24.1	377
397	Common variants at 7p21 are associated with frontotemporal lobar degeneration with TDP-43 inclusions. *Nature Genetics*, **2010**, 42, 234-9	36.3	361
396	Genome-wide association analyses identify new risk variants and the genetic architecture of amyotrophic lateral sclerosis. *Nature Genetics*, **2016**, 48, 1043-8	36.3	328
395	Familial amyotrophic lateral sclerosis-linked SOD1 mutants perturb fast axonal transport to reduce axonal mitochondria content. *Human Molecular Genetics*, **2007**, 16, 2720-2728	5.6	312
394	Oxidative stress in ALS: a mechanism of neurodegeneration and a therapeutic target. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **2006**, 1762, 1051-67	6.9	305
393	Genome-wide Analyses Identify KIF5A as a Novel ALS Gene. *Neuron*, **2018**, 97, 1268-1283.e6	13.9	296
392	Oxidative damage to protein in sporadic motor neuron disease spinal cord. *Annals of Neurology*, **1995**, 38, 691-5	9.4	282
391	Mitochondrial enzyme activity in amyotrophic lateral sclerosis: implications for the role of mitochondria in neuronal cell death. *Annals of Neurology*, **1999**, 46, 787-90	9.4	264
390	Update on the glutamatergic neurotransmitter system and the role of excitotoxicity in amyotrophic lateral sclerosis. *Muscle and Nerve*, **2002**, 26, 438-58	3.4	246

389	Astrocyte phenotype in relation to Alzheimer-type pathology in the ageing brain. *Neurobiology of Aging*, **2010**, 31, 578-90	5.6	243
388	The C9orf72 protein interacts with Rab1a and the ULK1 complex to regulate initiation of autophagy. *EMBO Journal*, **2016**, 35, 1656-76	13	237
387	Clinico-pathological features in amyotrophic lateral sclerosis with expansions in C9ORF72. *Brain*, **2012**, 135, 751-64	11.2	236
386	Direct conversion of patient fibroblasts demonstrates non-cell autonomous toxicity of astrocytes to motor neurons in familial and sporadic ALS. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 829-32	11.5	231
385	Exome-wide rare variant analysis identifies TUBA4A mutations associated with familial ALS. *Neuron*, **2014**, 84, 324-31	13.9	229
384	Parvalbumin and calbindin D-28k in the human motor system and in motor neuron disease. *Neuropathology and Applied Neurobiology*, **1993**, 19, 291-9	5.2	219
383	Systemic delivery of scAAV9 expressing SMN prolongs survival in a model of spinal muscular atrophy. *Science Translational Medicine*, **2010**, 2, 35ra42	17.5	210
382	Sequestration of multiple RNA recognition motif-containing proteins by C9orf72 repeat expansions. *Brain*, **2014**, 137, 2040-51	11.2	207
381	Mitochondrial dysfunction in a cell culture model of familial amyotrophic lateral sclerosis. *Brain*, **2002**, 125, 1522-33	11.2	205
380	The role of mitochondria in amyotrophic lateral sclerosis. *Neuroscience Letters*, **2019**, 710, 132933	3.3	191
379	Prognosis for patients with amyotrophic lateral sclerosis: development and validation of a personalised prediction model. *Lancet Neurology, The*, **2018**, 17, 423-433	24.1	189
378	CSF and plasma amino acid levels in motor neuron disease: elevation of CSF glutamate in a subset of patients. *Experimental Neurology*, **1995**, 4, 209-16		188
377	Calcium-permeable alpha-amino-3-hydroxy-5-methyl-4-isoxazole propionic acid receptors: a molecular determinant of selective vulnerability in amyotrophic lateral sclerosis. *Annals of Neurology*, **1997**, 42, 200-7	9.4	182
376	Chromosome 9p21 in sporadic amyotrophic lateral sclerosis in the UK and seven other countries: a genome-wide association study. *Lancet Neurology, The*, **2010**, 9, 986-94	24.1	171
375	Molecular factors underlying selective vulnerability of motor neurons to neurodegeneration in amyotrophic lateral sclerosis. *Journal of Neurology*, **2000**, 247 Suppl 1, I17-27	5.5	170
374	Mutations in CHMP2B in lower motor neuron predominant amyotrophic lateral sclerosis (ALS). *PLoS ONE*, **2010**, 5, e9872	3.7	170
373	Microglia as potential contributors to motor neuron injury in amyotrophic lateral sclerosis. *Glia*, **2005**, 51, 241-53	9	169
372	Amyotrophic lateral sclerosis: current issues in classification, pathogenesis and molecular pathology. *Neuropathology and Applied Neurobiology*, **1998**, 24, 104-17	5.2	165

371	Microarray analysis of the cellular pathways involved in the adaptation to and progression of motor neuron injury in the SOD1 G93A mouse model of familial ALS. *Journal of Neuroscience*, **2007**, 27, 9201-19	6.6	160
370	Detection of long repeat expansions from PCR-free whole-genome sequence data. *Genome Research*, **2017**, 27, 1895-1903	9.7	159
369	The C9ORF72 expansion mutation is a common cause of ALS+/-FTD in Europe and has a single founder. *European Journal of Human Genetics*, **2013**, 21, 102-8	5.3	159
368	Mitochondrial involvement in amyotrophic lateral sclerosis. *Neurochemistry International*, **2002**, 40, 543-51	4.4	159
367	Oxidative stress and motor neurone disease. *Brain Pathology*, **1999**, 9, 165-86	6	156
366	Convergent cholinergic activities in aging and Alzheimer's disease. *Neurobiology of Aging*, **1992**, 13, 393-400	3.6	156
365	Gene expression profiling in human neurodegenerative disease. *Nature Reviews Neurology*, **2012**, 8, 518-30	15	152
364	NEK1 variants confer susceptibility to amyotrophic lateral sclerosis. *Nature Genetics*, **2016**, 48, 1037-42	36.3	149
363	Mutant SOD1 alters the motor neuronal transcriptome: implications for familial ALS. *Brain*, **2005**, 128, 1686-706	11.2	148
362	Novel insertion in the KSP region of the neurofilament heavy gene in amyotrophic lateral sclerosis (ALS). *NeuroReport*, **1998**, 9, 3967-70	1.7	142
361	Phase 1-2 Trial of Antisense Oligonucleotide Tofersen for ALS. *New England Journal of Medicine*, **2020**, 383, 109-119	59.2	140
360	Neuronal dark matter: the emerging role of microRNAs in neurodegeneration. *Frontiers in Cellular Neuroscience*, **2013**, 7, 178	6.1	135
359	Diagnosis and management of motor neurone disease. *BMJ, The*, **2008**, 336, 658-62	5.9	134
358	Microarray analysis of the astrocyte transcriptome in the aging brain: relationship to Alzheimer's pathology and APOE genotype. *Neurobiology of Aging*, **2011**, 32, 1795-807	5.6	133
357	White matter lesions in an unselected cohort of the elderly: astrocytic, microglial and oligodendrocyte precursor cell responses. *Neuropathology and Applied Neurobiology*, **2007**, 33, 410-9	5.2	129
356	Dysregulation of astrocyte-motoneuron cross-talk in mutant superoxide dismutase 1-related amyotrophic lateral sclerosis. *Brain*, **2011**, 134, 2627-41	11.2	127
355	The role of TREM2 R47H as a risk factor for Alzheimer's disease, frontotemporal lobar degeneration, amyotrophic lateral sclerosis, and Parkinson's disease. *Alzheimerjs and Dementia*, **2015**, 11, 1407-1416	1.2	126
354	The widening spectrum of C9ORF72-related disease; genotype/phenotype correlations and potential modifiers of clinical phenotype. *Acta Neuropathologica*, **2014**, 127, 333-45	14.3	124

353	Astrocyte function and role in motor neuron disease: a future therapeutic target?. *Glia*, **2009**, 57, 1251-64	9	124
352	Molecular pathology and genetic advances in amyotrophic lateral sclerosis: an emerging molecular pathway and the significance of glial pathology. *Acta Neuropathologica*, **2011**, 122, 657-71	14.3	120
351	Antisense RNA foci in the motor neurons of C9ORF72-ALS patients are associated with TDP-43 proteinopathy. *Acta Neuropathologica*, **2015**, 130, 63-75	14.3	118
350	Apoptosis in amyotrophic lateral sclerosis: a review of the evidence. *Neuropathology and Applied Neurobiology*, **2001**, 27, 257-74	5.2	118
349	Mechanisms, models and biomarkers in amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2013**, 14 Suppl 1, 19-32	3.6	114
348	The RNA of the glutamate transporter EAAT2 is variably spliced in amyotrophic lateral sclerosis and normal individuals. *Journal of the Neurological Sciences*, **1999**, 170, 45-50	3.2	113
347	Protein aggregation in motor neurone disorders. *Neuropathology and Applied Neurobiology*, **2003**, 29, 529-45	5.2	112
346	The microtubule-severing protein Spastin is essential for axon outgrowth in the zebrafish embryo. *Human Molecular Genetics*, **2006**, 15, 2763-71	5.6	110
345	Familial amyotrophic lateral sclerosis with a mutation in exon 4 of the Cu/Zn superoxide dismutase gene: pathological and immunocytochemical changes. *Acta Neuropathologica*, **1996**, 92, 395-403	14.3	110
344	Early involvement of the spinal cord in diabetic peripheral neuropathy. *Diabetes Care*, **2006**, 29, 2664-9	14.6	108
343	Secondary reduction in calpain 3 expression in patients with limb girdle muscular dystrophy type 2B and Miyoshi myopathy (primary dysferlinopathies). *Neuromuscular Disorders*, **2000**, 10, 553-9	2.9	106
342	Direct evidence for axonal transport defects in a novel mouse model of mutant spastin-induced hereditary spastic paraplegia (HSP) and human HSP patients. *Journal of Neurochemistry*, **2009**, 110, 34-44	6	104
341	Chromosome 9 ALS and FTD locus is probably derived from a single founder. *Neurobiology of Aging*, **2012**, 33, 209.e3-8	5.6	103
340	ALS-associated mutations in FUS disrupt the axonal distribution and function of SMN. *Human Molecular Genetics*, **2013**, 22, 3690-704	5.6	103
339	Oligodendrocytes contribute to motor neuron death in ALS via SOD1-dependent mechanism. *Proceedings of the National Academy of Sciences of the United States of America*, **2016**, 113, E6496-E6505	11.5	102
338	Novel FUS/TLS mutations and pathology in familial and sporadic amyotrophic lateral sclerosis. *Archives of Neurology*, **2010**, 67, 455-61		99
337	Hereditary spastic paraparesis: disrupted intracellular transport associated with spastin mutation. *Annals of Neurology*, **2003**, 54, 748-59	9.4	98
336	TMEM106B is a genetic modifier of frontotemporal lobar degeneration with C9orf72 hexanucleotide repeat expansions. *Acta Neuropathologica*, **2014**, 127, 407-18	14.3	97

335	PTEN depletion rescues axonal growth defect and improves survival in SMN-deficient motor neurons. *Human Molecular Genetics*, **2010**, 19, 3159-68	5.6	97
334	The expression of the glial glutamate transporter protein EAAT2 in motor neuron disease: an immunohistochemical study. *European Journal of Neuroscience*, **1998**, 10, 2481-9	3.5	97
333	Unravelling the enigma of selective vulnerability in neurodegeneration: motor neurons resistant to degeneration in ALS show distinct gene expression characteristics and decreased susceptibility to excitotoxicity. *Acta Neuropathologica*, **2013**, 125, 95-109	14.3	96
332	The expression of neuronal voltage-dependent calcium channels in human cerebellum. *Molecular Brain Research*, **1995**, 34, 271-82		96
331	A genome-wide association meta-analysis identifies a novel locus at 17q11.2 associated with sporadic amyotrophic lateral sclerosis. *Human Molecular Genetics*, **2014**, 23, 2220-31	5.6	95
330	LRP-1-mediated intracellular antibody delivery to the Central Nervous System. *Scientific Reports*, **2015**, 5, 11990	4.9	95
329	Amyotrophic lateral sclerosis associated with genetic abnormalities in the gene encoding Cu/Zn superoxide dismutase: molecular pathology of five new cases, and comparison with previous reports and 73 sporadic cases of ALS. *Journal of Neuropathology and Experimental Neurology*, **1998**, 57, 895-904	3.1	95
328	Lithium in patients with amyotrophic lateral sclerosis (LiCALS): a phase 3 multicentre, randomised, double-blind, placebo-controlled trial. *Lancet Neurology, The*, **2013**, 12, 339-45	24.1	94
327	Expression of the glial glutamate transporter EAAT2 in the human CNS: an immunohistochemical study. *Molecular Brain Research*, **1997**, 52, 17-31		94
326	Development and characterisation of a glutamate-sensitive motor neurone cell line. *Journal of Neurochemistry*, **2000**, 74, 1895-902	6	92
325	Iron, selenium and glutathione peroxidase activity are elevated in sporadic motor neuron disease. *Neuroscience Letters*, **1994**, 182, 87-90	3.3	91
324	C9orf72 expansion disrupts ATM-mediated chromosomal break repair. *Nature Neuroscience*, **2017**, 20, 1225-1235	25.5	90
323	Microglial activation in white matter lesions and nonlesional white matter of ageing brains. *Neuropathology and Applied Neurobiology*, **2007**, 33, 670-83	5.2	88
322	Production of monocyte chemoattractant protein-1 in amyotrophic lateral sclerosis. *Muscle and Nerve*, **2005**, 32, 541-4	3.4	88
321	C9ORF72 interaction with cofilin modulates actin dynamics in motor neurons. *Nature Neuroscience*, **2016**, 19, 1610-1618	25.5	87
320	Vascular endothelial growth factor and the nervous system. *Neuropathology and Applied Neurobiology*, **2004**, 30, 427-46	5.2	86
319	Analysis of the cytosolic proteome in a cell culture model of familial amyotrophic lateral sclerosis reveals alterations to the proteasome, antioxidant defenses, and nitric oxide synthetic pathways. *Journal of Biological Chemistry*, **2003**, 278, 6371-83	5.4	85
318	A phase II-III trial of olesoxime in subjects with amyotrophic lateral sclerosis. *European Journal of Neurology*, **2014**, 21, 529-36	6	83

317	Rasch analysis of the hospital anxiety and depression scale (HADS) for use in motor neurone disease. *Health and Quality of Life Outcomes*, **2011**, 9, 82	3	79
316	Excitotoxicity and motor neurone disease: a review of the evidence. *Journal of the Neurological Sciences*, **1994**, 124 Suppl, 6-13	3.2	79
315	Peroxynitrite and hydrogen peroxide induced cell death in the NSC34 neuroblastoma x spinal cord cell line: role of poly (ADP-ribose) polymerase. *Journal of Neurochemistry*, **1998**, 70, 501-8	6	78
314	The cellular and molecular pathology of the motor system in hereditary spastic paraparesis due to mutation of the spastin gene. *Journal of Neuropathology and Experimental Neurology*, **2003**, 62, 1166-77	3.1	77
313	Project MinE: study design and pilot analyses of a large-scale whole-genome sequencing study in amyotrophic lateral sclerosis. *European Journal of Human Genetics*, **2018**, 26, 1537-1546	5.3	75
312	Expression of vascular endothelial growth factor and its receptors in the central nervous system in amyotrophic lateral sclerosis. *Journal of Neuropathology and Experimental Neurology*, **2006**, 65, 26-36	3.1	75
311	Serum miRNAs miR-206, 143-3p and 374b-5p as potential biomarkers for amyotrophic lateral sclerosis (ALS). *Neurobiology of Aging*, **2017**, 55, 123-131	5.6	74
310	Mutations in the vesicular trafficking protein annexin A11 are associated with amyotrophic lateral sclerosis. *Science Translational Medicine*, **2017**, 9,	17.5	74
309	C9ORF72 GGGGCC Expanded Repeats Produce Splicing Dysregulation which Correlates with Disease Severity in Amyotrophic Lateral Sclerosis. *PLoS ONE*, **2015**, 10, e0127376	3.7	74
308	The quantitative autoradiographic distribution of [3H]MK-801 binding sites in the normal human spinal cord. *Brain Research*, **1991**, 539, 164-8	3.7	74
307	Loss of nuclear TDP-43 in amyotrophic lateral sclerosis (ALS) causes altered expression of splicing machinery and widespread dysregulation of RNA splicing in motor neurones. *Neuropathology and Applied Neurobiology*, **2014**, 40, 670-85	5.2	73
306	Alterations in the blood brain barrier in ageing cerebral cortex in relationship to Alzheimer-type pathology: a study in the MRC-CFAS population neuropathology cohort. *Neuroscience Letters*, **2011**, 505, 25-30	3.3	73
305	Shared polygenic risk and causal inferences in amyotrophic lateral sclerosis. *Annals of Neurology*, **2019**, 85, 470-481	9.4	72
304	SRSF1-dependent nuclear export inhibition of C9ORF72 repeat transcripts prevents neurodegeneration and associated motor deficits. *Nature Communications*, **2017**, 8, 16063	17.4	71
303	Population variation in oxidative stress and astrocyte DNA damage in relation to Alzheimer-type pathology in the ageing brain. *Neuropathology and Applied Neurobiology*, **2010**, 36, 25-40	5.2	71
302	Neuro-ophthalmological complications of coronary artery bypass graft surgery. *Acta Neurologica Scandinavica*, **1987**, 76, 1-7	3.8	71
301	An in vitro screening cascade to identify neuroprotective antioxidants in ALS. *Free Radical Biology and Medicine*, **2009**, 46, 1127-38	7.8	70
300	Broad clinical phenotypes associated with TAR-DNA binding protein (TARDBP) mutations in amyotrophic lateral sclerosis. *Neurogenetics*, **2010**, 11, 217-25	3	68

299	Differential localization of voltage-dependent calcium channel alpha1 subunits at the human and rat neuromuscular junction. *Journal of Neuroscience*, **1997**, 17, 6226-35	6.6	67
298	Apoptosis in amyotrophic lateral sclerosis--what is the evidence?. *Lancet Neurology, The*, **2005**, 4, 500-9	24.1	66
297	Roadmap and standard operating procedures for biobanking and discovery of neurochemical markers in ALS. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders*, **2012**, 13, 1-10		65
296	Early interneuron dysfunction in ALS: insights from a mutant sod1 zebrafish model. *Annals of Neurology*, **2013**, 73, 246-58	9.4	65
295	Distribution of alpha 1A, alpha 1B and alpha 1E voltage-dependent calcium channel subunits in the human hippocampus and parahippocampal gyrus. *Neuroscience*, **1996**, 71, 1013-24	3.9	65
294	Multicenter validation of CSF neurofilaments as diagnostic biomarkers for ALS. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2016**, 17, 404-13	3.6	65
293	Safety and efficacy of diaphragm pacing in patients with respiratory insufficiency due to amyotrophic lateral sclerosis (DiPALS): a multicentre, open-label, randomised controlled trial. *Lancet Neurology, The*, **2015**, 14, 883-892	24.1	64
292	Optimised and rapid pre-clinical screening in the SOD1(G93A) transgenic mouse model of amyotrophic lateral sclerosis (ALS). *PLoS ONE*, **2011**, 6, e23244	3.7	64
291	Review: The role of mitochondria in the pathogenesis of amyotrophic lateral sclerosis. *Neuropathology and Applied Neurobiology*, **2011**, 37, 336-52	5.2	64
290	Meta-analysis of vascular endothelial growth factor variations in amyotrophic lateral sclerosis: increased susceptibility in male carriers of the -2578AA genotype. *Journal of Medical Genetics*, **2009**, 46, 840-6	5.8	63
289	Small RNA Sequencing of Sporadic Amyotrophic Lateral Sclerosis Cerebrospinal Fluid Reveals Differentially Expressed miRNAs Related to Neural and Glial Activity. *Frontiers in Neuroscience*, **2017**, 11, 731	5.1	62
288	[3H]D-aspartate binding sites in the normal human spinal cord and changes in motor neuron disease: a quantitative autoradiographic study. *Brain Research*, **1994**, 655, 195-201	3.7	62
287	C9orf72 and UNC13A are shared risk loci for amyotrophic lateral sclerosis and frontotemporal dementia: a genome-wide meta-analysis. *Annals of Neurology*, **2014**, 76, 120-33	9.4	61
286	Non-invasive ventilation in motor neuron disease: an update of current UK practice. *Journal of Neurology, Neurosurgery and Psychiatry*, **2012**, 83, 371-6	5.5	61
285	Thalamic neuronal dysfunction and chronic sensorimotor distal symmetrical polyneuropathy in patients with type 1 diabetes mellitus. *Diabetologia*, **2008**, 51, 2088-92	10.3	61
284	Physical activity as an exogenous risk factor in motor neuron disease (MND): a review of the evidence. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders*, **2009**, 10, 191-204		60
283	Selective loss of neurofilament expression in Cu/Zn superoxide dismutase (SOD1) linked amyotrophic lateral sclerosis. *Journal of Neurochemistry*, **2002**, 82, 1118-28	6	60
282	Mood disturbances in motor neurone disease. *Journal of the Neurological Sciences*, **1998**, 160 Suppl 1, S53-6	3.2	60

281	Alterations of the blood-brain barrier in cerebral white matter lesions in the ageing brain. *Neuroscience Letters*, **2010**, 486, 246-51	3.3	59
280	Microarray RNA expression analysis of cerebral white matter lesions reveals changes in multiple functional pathways. *Stroke*, **2009**, 40, 369-75	6.7	59
279	Use of clinical staging in amyotrophic lateral sclerosis for phase 3 clinical trials. *Journal of Neurology, Neurosurgery and Psychiatry*, **2015**, 86, 45-9	5.5	58
278	Phosphatase and tensin homologue/protein kinase B pathway linked to motor neuron survival in human superoxide dismutase 1-related amyotrophic lateral sclerosis. *Brain*, **2011**, 134, 506-17	11.2	57
277	Exome sequencing in amyotrophic lateral sclerosis implicates a novel gene, DNAJC7, encoding a heat-shock protein. *Nature Neuroscience*, **2019**, 22, 1966-1974	25.5	56
276	The El Escorial criteria: strengths and weaknesses. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2015**, 16, 1-7	3.6	55
275	Multicentre quality control evaluation of different biomarker candidates for amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2014**, 15, 344-50	3.6	55
274	Impairment of mitochondrial anti-oxidant defence in SOD1-related motor neuron injury and amelioration by ebselen. *Brain*, **2006**, 129, 1693-709	11.2	55
273	Transcriptomic indices of fast and slow disease progression in two mouse models of amyotrophic lateral sclerosis. *Brain*, **2013**, 136, 3305-32	11.2	54
272	The natural history of motor neuron disease: assessing the impact of specialist care. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2013**, 14, 13-9	3.6	54
271	Oxidative damage and motor neurone disease difficulties in the measurement of protein carbonyls in human brain tissue. *Free Radical Research*, **1996**, 24, 397-406	4	53
270	Glial proliferation and metabotropic glutamate receptor expression in amyotrophic lateral sclerosis. *Journal of Neuropathology and Experimental Neurology*, **2004**, 63, 831-40	3.1	53
269	Biomarkers in Motor Neuron Disease: A State of the Art Review. *Frontiers in Neurology*, **2019**, 10, 291	4.1	52
268	Superoxide dismutase 1 mutation in a cellular model of amyotrophic lateral sclerosis shifts energy generation from oxidative phosphorylation to glycolysis. *Neurobiology of Aging*, **2014**, 35, 1499-509	5.6	51
267	ANO5 gene analysis in a large cohort of patients with anoctaminopathy: confirmation of male prevalence and high occurrence of the common exon 5 gene mutation. *Human Mutation*, **2013**, 34, 1111-8	4.7	51
266	Motor neuron disease in a patient with a mitochondrial tRNAIle mutation. *Annals of Neurology*, **2006**, 59, 570-4	9.4	50
265	C9ORF72 expansions, parkinsonism, and Parkinson disease: a clinicopathologic study. *Neurology*, **2013**, 81, 808-11	6.5	49
264	Vascular endothelial growth factor counteracts the loss of phospho-Akt preceding motor neurone degeneration in amyotrophic lateral sclerosis. *Neuropathology and Applied Neurobiology*, **2007**, 33, 499-509	5.2	49

263	Low expression of GluR2 AMPA receptor subunit protein by human motor neurons. *NeuroReport*, **1999**, 10, 261-5	1.7	49
262	Meta-analysis of pharmacogenetic interactions in amyotrophic lateral sclerosis clinical trials. *Neurology*, **2017**, 89, 1915-1922	6.5	48
261	Pattern of spread and prognosis in lower limb-onset ALS. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders*, **2010**, 11, 369-73		47
260	A comprehensive analysis of rare genetic variation in amyotrophic lateral sclerosis in the UK. *Brain*, **2017**, 140, 1611-1618	11.2	46
259	S[+] Apomorphine is a CNS penetrating activator of the Nrf2-ARE pathway with activity in mouse and patient fibroblast models of amyotrophic lateral sclerosis. *Free Radical Biology and Medicine*, **2013**, 61, 438-52	7.8	46
258	Concurrence of multiple sclerosis and amyotrophic lateral sclerosis in patients with hexanucleotide repeat expansions of C9ORF72. *Journal of Neurology, Neurosurgery and Psychiatry*, **2013**, 84, 79-87	5.5	46
257	Gene expression signatures in motor neurone disease fibroblasts reveal dysregulation of metabolism, hypoxia-response and RNA processing functions. *Neuropathology and Applied Neurobiology*, **2015**, 41, 201-26	5.2	45
256	Nonverbal visual attention, but not recognition memory of learning, processes are impaired in motor neurone disease. *Neuropsychologia*, **1996**, 34, 377-85	3.2	45
255	Astrocyte adenosine deaminase loss increases motor neuron toxicity in amyotrophic lateral sclerosis. *Brain*, **2019**, 142, 586-605	11.2	44
254	Amyotrophic lateral sclerosis: a consensus viewpoint on designing and implementing a clinical trial. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders: Official Publication of the World Federation of Neurology, Research Group on Motor Neuron Diseases*, **2004**, 5, 84-98		44
253	Immune response in peripheral axons delays disease progression in SOD1 mice. *Journal of Neuroinflammation*, **2016**, 13, 261	10.1	44
252	ALS-associated missense and nonsense TBK1 mutations can both cause loss of kinase function. *Neurobiology of Aging*, **2018**, 71, 266.e1-266.e10	5.6	44
251	Differences in protein quality control correlate with phenotype variability in 2 mouse models of familial amyotrophic lateral sclerosis. *Neurobiology of Aging*, **2015**, 36, 492-504	5.6	43
250	A new zebrafish model produced by TILLING of SOD1-related amyotrophic lateral sclerosis replicates key features of the disease and represents a tool for in vivo therapeutic screening. *DMM Disease Models and Mechanisms*, **2014**, 7, 73-81	4.1	43
249	Gastrostomy use in motor neurone disease (MND): a review, meta-analysis and survey of current practice. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2013**, 14, 96-104	3.6	43
248	Cu/Zn superoxide dismutase (SOD1) mutations associated with familial amyotrophic lateral sclerosis (ALS) affect cellular free radical release in the presence of oxidative stress. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders: Official Publication of the World Federation of Neurology, Research Group on Motor Neuron Diseases*, **2002**, 3, 75-85		43
247	An immunocytochemical study of the distribution of AMPA selective glutamate receptor subunits in the normal human motor system. *Neuroscience*, **1996**, 74, 185-98	3.9	43
246	Population variation in glial fibrillary acidic protein levels in brain ageing: relationship to Alzheimer-type pathology and dementia. *Dementia and Geriatric Cognitive Disorders*, **2009**, 27, 465-73	2.6	41

245	PTEN depletion decreases disease severity and modestly prolongs survival in a mouse model of spinal muscular atrophy. *Molecular Therapy*, **2015**, 23, 270-7	11.7	40
244	Association of a Locus in the CAMTA1 Gene With Survival in Patients With Sporadic Amyotrophic Lateral Sclerosis. *JAMA Neurology*, **2016**, 73, 812-20	17.2	40
243	Simultaneous and independent detection of C9ORF72 alleles with low and high number of GGGGCC repeats using an optimised protocol of Southern blot hybridisation. *Molecular Neurodegeneration*, **2013**, 8, 12	19	40
242	The Spectrum of C9orf72-mediated Neurodegeneration and Amyotrophic Lateral Sclerosis. *Neurotherapeutics*, **2015**, 12, 326-39	6.4	39
241	Motor neurone disease: a practical update on diagnosis and management. *Clinical Medicine*, **2010**, 10, 252-8	1.9	39
240	A data-driven approach links microglia to pathology and prognosis in amyotrophic lateral sclerosis. *Acta Neuropathologica Communications*, **2017**, 5, 23	7.3	38
239	Non-NMDA receptors in motor neuron disease (MND): a quantitative autoradiographic study in spinal cord and motor cortex using [3H]CNQX and [3H]kainate. *Brain Research*, **1994**, 655, 186-94	3.7	38
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