Adrian M Isaacs

List of Publications by Year
in descending order

84	11,570	57758 44	64796 79
papers	citations	h-index	g-index

101	101	101	11833
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	Association of missense and 5â€²-splice-site mutations in tau with the inherited dementia FTDP-17. Nature, 1998, 393, 702-705.	27.8	3,333
2	A systematic, genome-wide, phenotype-driven mutagenesis programme for gene function studies in the mouse. Nature Genetics, 2000, 25, 440-443.	21.4	657
3	C9orf72 repeat expansions cause neurodegeneration in Drosophila through arginine-rich proteins. Science, 2014, 345, 1192-1194.	12.6	632
4	Functional multivesicular bodies are required for autophagic clearance of protein aggregates associated with neurodegenerative disease. Journal of Cell Biology, 2007, 179, 485-500.	5.2	559
5	C9orf72-mediated ALS and FTD: multiple pathways to disease. Nature Reviews Neurology, 2018, 14, 544-558.	10.1	478
6	Clinical and neuroanatomical signatures of tissue pathology in frontotemporal lobar degeneration. Brain, 2011, 134, 2565-2581.	7.6	306
7	Large C9orf72 Hexanucleotide Repeat Expansions Are Seen in Multiple Neurodegenerative Syndromes and Are More Frequent Than Expected in the UK Population. American Journal of Human Genetics, 2013, 92, 345-353.	6.2	297
8	C9orf72 frontotemporal lobar degeneration is characterised by frequent neuronal sense and antisense RNA foci. Acta Neuropathologica, 2013, 126, 845-857.	7.7	289
9	Evolution of the Aging Brain Transcriptome and Synaptic Regulation. PLoS ONE, 2008, 3, e3329.	2.5	285
10	C9orf72 hexanucleotide repeat associated with amyotrophic lateral sclerosis and frontotemporal dementia forms RNA G-quadruplexes. Scientific Reports, 2012, 2, 1016.	3.3	275
11	C9orf72 expansions in frontotemporal dementia and amyotrophic lateral sclerosis. Lancet Neurology, The, 2015, 14, 291-301.	10.2	270
12	Conformation Determines the Seeding Potencies of Native and Recombinant Tau Aggregates. Journal of Biological Chemistry, 2015, 290, 1049-1065.	3.4	225
13	A distinct clinical, neuropsychological and radiological phenotype is associated with progranulin gene mutations in a large UK series. Brain, 2008, 131, 706-720.	7.6	222
14	FUS pathology defines the majority of tau- and TDP-43-negative frontotemporal lobar degeneration. Acta Neuropathologica, 2010, 120, 33-41.	7.7	222
15	Gâ€quadruplexes: Emerging roles in neurodegenerative diseases and the nonâ€coding transcriptome. FEBS Letters, 2015, 589, 1653-1668.	2.8	185
16	Gâ€quadruplexâ€binding small molecules ameliorate C9orf72 <scp>FTD</scp> / <scp>ALS</scp> pathology inÂvitro and inÂvivo. EMBO Molecular Medicine, 2018, 10, 22-31.	6.9	178
17	Disruption of endocytic trafficking in frontotemporal dementia with CHMP2B mutations. Human Molecular Genetics, 2010, 19, 2228-2238.	2.9	163
18	TDP-43 extracted from frontotemporal lobar degeneration subject brains displays distinct aggregate assemblies and neurotoxic effects reflecting disease progression rates. Nature Neuroscience, 2019, 22, 65-77.	14.8	143

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19	CHMP2B C-truncating mutations in frontotemporal lobar degeneration are associated with an aberrant endosomal phenotype in vitro. Human Molecular Genetics, 2008, 17, 313-322.	2.9	131
20	Mice with endogenous <scp>TDP</scp> â€43 mutations exhibit gain of splicing function and characteristics of amyotrophic lateral sclerosis. EMBO Journal, 2018, 37, .	7.8	129
21	Homozygosity for the C9orf72 GGGGCC repeat expansion in frontotemporal dementia. Acta Neuropathologica, 2013, 126, 401-409.	7.7	126
22	Acceleration of Amyloid Î²-Peptide Aggregation by Physiological Concentrations of Calcium. Journal of Biological Chemistry, 2006, 281, 27916-27923.	3.4	125
23	A Reassessment of the Neuropathology of Frontotemporal Dementia Linked to Chromosome 3. Journal of Neuropathology and Experimental Neurology, 2007, 66, 884-891.	1.7	118
24	SRSF1-dependent nuclear export inhibition of C9ORF72 repeat transcripts prevents neurodegeneration and associated motor deficits. Nature Communications, 2017, 8, 16063.	12.8	106
25	A zebrafish model for C9orf72 ALS reveals RNA toxicity as a pathogenic mechanism. Acta Neuropathologica, 2018, 135, 427-443.	7.7	98
26	Progressive neuronal inclusion formation and axonal degeneration in CHMP2B mutant transgenic mice. Brain, 2012, 135, 819-832.	7.6	97
27	Humanized mutant FUS drives progressive motor neuron degeneration without aggregation in â€˜FUSDelta14â€™ knockin mice. Brain, 2017, 140, 2797-2805.	7.6	95
28	C9orf72 arginine-rich dipeptide proteins interact with ribosomal proteins in vivo to induce a toxic translational arrest that is rescued by eIF1A. Acta Neuropathologica, 2019, 137, 487-500.	7.7	94
29	RPS25 is required for efficient RAN translation of C9orf72 and other neurodegenerative disease-associated nucleotide repeats. Nature Neuroscience, 2019, 22, 1383-1388.	14.8	87
30	Localization of frontotemporal dementia with parkinsonism in an Australian kindred to chromosome 17q21-22. Annals of Neurology, 1997, 42, 794-798.	5.3	83
31	Quantitative analysis of cryptic splicing associated with TDP-43 depletion. BMC Medical Genomics, 2017, 10, 38.	1.5	81
32	Frontotemporal dementia caused by CHMP2B mutation is characterised by neuronal lysosomal storage pathology. Acta Neuropathologica, 2015, 130, 511-523.	7.7	79
33	A comparative clinical, pathological, biochemical and genetic study of fused in sarcoma proteinopathies. Brain, 2011, 134, 2548-2564.	7.6	76
34	FUS ALS-causative mutations impair FUS autoregulation and splicing factor networks through intron retention. Nucleic Acids Research, 2020, 48, 6889-6905.	14.5	70
35	The role of ESCRT proteins in fusion events involving lysosomes, endosomes and autophagosomes. Biochemical Society Transactions, 2010, 38, 1469-1473.	3.4	69
36	C9orf72 amyotrophic lateral sclerosis and frontotemporal dementia. Current Opinion in Neurology, 2014, 27, 515-523.	3.6	67

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37	Quantitative Assessment of Eye Phenotypes for Functional Genetic Studies UsingDrosophila melanogaster. G3: Genes, Genomes, Genetics, 2016, 6, 1427-1437.	1.8	67
38	A Mutation inAf4Is Predicted to Cause Cerebellar Ataxia and Cataracts in the Robotic Mouse. Journal of Neuroscience, 2003, 23, 1631-1637.	3.6	66
39	TDP-43 is a culprit in human neurodegeneration, and not just an innocent bystander. Mammalian Genome, 2008, 19, 299-305.	2.2	63
40	Ageing as a risk factor for ALS/FTD. Human Molecular Genetics, 2017, 26, R105-R113.	2.9	61
41	The role of CHMP2B in frontotemporal dementia. Biochemical Society Transactions, 2009, 37, 208-212.	3.4	60
42	Sense and antisense RNA are not toxic in Drosophila models of C9orf72-associated ALS/FTD. Acta Neuropathologica, 2018, 135, 445-457.	7.7	59
43	Absence of FUS-immunoreactive pathology in frontotemporal dementia linked to chromosome 3 (FTD-3) caused by mutation in the CHMP2B gene. Acta Neuropathologica, 2009, 118, 719-720.	7.7	56
44	Regulation of Postsynaptic Function by the Dementia-Related ESCRT-III Subunit CHMP2B. Journal of Neuroscience, 2015, 35, 3155-3173.	3.6	50
45	Genetic models of C9orf72: what is toxic?. Current Opinion in Genetics and Development, 2017, 44, 92-101.	3.3	50
46	Bidirectional nucleolar dysfunction in C9orf72 frontotemporal lobar degeneration. Acta Neuropathologica Communications, 2017, 5, 29.	5.2	43
47	Symmetric dimethylation of poly-GR correlates with disease duration in C9orf72 FTLD and ALS and reduces poly-GR phase separation and toxicity. Acta Neuropathologica, 2020, 139, 407-410.	7.7	36
48	Identification of a New Pmp22 Mouse Mutant and Trafficking Analysis of a Pmp22 Allelic Series Suggesting That Protein Aggregates May Be Protective in Pmp22-Associated Peripheral Neuropathy. Molecular and Cellular Neurosciences, 2002, 21, 114-125.	2.2	34
49	Novel Types of Frontotemporal Lobar Degeneration: Beyond Tau and TDP-43. Journal of Molecular Neuroscience, 2011, 45, 402-408.	2.3	33
50	A 30-unit hexanucleotide repeat expansion in C9orf72 induces pathological lesions with dipeptide-repeat proteins and RNA foci, but not TDP-43 inclusions and clinical disease. Acta Neuropathologica, 2015, 130, 599-601.	7.7	31
51	A novel exon 2 I27V VCP variant is associated with dissimilar clinical syndromes. Journal of Neurology, 2011, 258, 1494-1496.	3.6	30
52	Cortical volumes and atrophy rates in FTD-3 CHMP2B mutation carriers and related non-carriers. NeuroImage, 2009, 45, 713-721.	4.2	28
53	Frontotemporal dementia causative CHMP2B impairs neuronal endolysosomal traffic-rescue byTMEM106Bknockdown. Brain, 2018, 141, 3428-3442.	7.6	27
54	Heterogeneous nuclear ribonucleoproteins R and Q accumulate in pathological inclusions in FTLD-FUS. Acta Neuropathologica Communications, 2019, 7, 18.	5.2	26

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55	Soluble and insoluble dipeptide repeat protein measurements in C9orf72-frontotemporal dementia brains show regional differential solubility and correlation of poly-GR with clinical severity. Acta Neuropathologica Communications, 2020, 8, 184.	5.2	25
56	Determination of the Gene Structure of Human GFAP and Absence of Coding Region Mutations Associated with Frontotemporal Dementia with Parkinsonism Linked to Chromosome 17. Genomics, 1998, 51, 152-154.	2.9	24
57	Early microgliosis precedes neuronal loss and behavioural impairment in mice with a frontotemporal dementia-causing CHMP2B mutation. Human Molecular Genetics, 2017, 26, ddx003.	2.9	22
58	RANTing about C9orf72. Neuron, 2013, 77, 597-598.	8.1	19
59	Enhanced insulin signalling ameliorates C9orf72 hexanucleotide repeat expansion toxicity in Drosophila. ELife, 2021, 10, .	6.0	18
60	Towards a mutant map of the mouse ? new models of neurological, behavioural, deafness, bone, renal and blood disorders. Genetica, 2004, 122, 47-49.	1.1	17
61	Presymptomatic Generalized Brain Atrophy in Frontotemporal Dementia Caused by CHMP2B Mutation. Dementia and Geriatric Cognitive Disorders, 2009, 27, 182-186.	1.5	17
62	Glycine-alanine dipeptide repeats spread rapidly in a repeat length- and age-dependent manner in the fly brain. Acta Neuropathologica Communications, 2019, 7, 209.	5.2	17
63	The integration site of the APP transgene in the J20 mouse model of Alzheimerâ€™s disease. Wellcome Open Research, 2017, 2, 84.	1.8	15
64	Reversal of pathology in CHMP2Bâ€mediated frontotemporal dementia patient cells using RNA interference. Journal of Gene Medicine, 2012, 14, 521-529.	2.8	14
65	The snowball effect of RNA binding protein dysfunction in amyotrophic lateral sclerosis. Brain, 2018, 141, 1236-1238.	7.6	14
66	Presymptomatic cerebral blood flow changes inCHMP2Bmutation carriers of familial frontotemporal dementia (FTD-3), measured with MRI. BMJ Open, 2012, 2, e000368.	1.9	13
67	C9orf72 ALS/FTD dipeptide repeat protein levels are reduced by small molecules that inhibit PKA or enhance protein degradation. EMBO Journal, 2022, 41, e105026.	7.8	13
68	Development of a sensitive trial-ready poly(GP) CSF biomarker assay forC9orf72-associated frontotemporal dementia and amyotrophic lateral sclerosis. Journal of Neurology, Neurosurgery and Psychiatry, 2022, 93, 761-771.	1.9	12
69	Specific biomarkers for C9orf72 <scp>FTD</scp>/<scp>ALS</scp> could expedite the journey towards effective therapies. EMBO Molecular Medicine, 2017, 9, 853-855.	6.9	10
70	Humoral response to neurofilaments and dipeptide repeats in ALS progression. Annals of Clinical and Translational Neurology, 2021, 8, 1831-1844.	3.7	8
71	Construction of a Detailed Physical and Transcript Map of the FTDP-17 Candidate Region on Chromosome 17q21. Genomics, 1999, 60, 129-136.	2.9	7
72	Adult polyglucosan body disease with <scp>GBE1</scp> haploinsufficiency and concomitant frontotemporal lobar degeneration. Neuropathology and Applied Neurobiology, 2014, 40, 778-782.	3.2	7

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73	Progranulin and TMEM106B: when two become wan. EMBO Reports, 2020, 21, e51668.	4.5	5
74	TMEM106B and ApoE polymorphisms in CHMP2B-mediated frontotemporal dementia (FTD-3). Neurobiology of Aging, 2017, 59, 221.e1-221.e7.	3.1	4
75	Six generations of CHMP2B â€mediated Frontotemporal Dementia: Clinical features, predictive testing, progression, and survival. Acta Neurologica Scandinavica, 2022, 145, 529-540.	2.1	4
76	A novel synaptopathyâ€defective synaptic vesicle protein trafficking in the mutant CHMP2B mouse model of frontotemporal dementia. Journal of Neurochemistry, 2022, 160, 412-425.	3.9	4
77	One target for amyotrophic lateral sclerosis therapy?. Science, 2016, 353, 647-648.	12.6	2
78	Relax, Donâ€™t RAN Translate It. Neuron, 2019, 104, 827-829.	8.1	2
79	Sizing, stabilising, and cloning repeat-expansions for gene targeting constructs. Methods, 2021, 191, 15-22.	3.8	2
80	P3-049: CHARACTERISATION OF A CO-CULTURE CELL-BASED MODEL OF TAU AGGREGATION AND PROPAGATION. , 2014, 10, P646-P646.		0
81	Molecular mechanisms and therapeutic strategies in amyotrophic lateral sclerosis caused by C9orf72 mutations. Lancet, The, 2016, 387, S13.	13.7	0
82	[P3â€“150]: SCREENING FOR MODIFIERS OF C9ORF72 HEXANUCLEOTIDE REPEAT EXPANSION TOXICITY IN DROSOPHILA. Alzheimer's and Dementia, 2017, 13, P992.	0.8	0
83	[S5â€“01â€“02]: TARGETING G4C2 Gâ€QUADRUPLEXES IN C9ORF72 FTD/ALS. Alzheimer's and Dementia, 2017, 13, P1444.	0.8	0
84	Haploinsufficiency of progranulin causes impairments in PINK/PARKIN mitophagy. Alzheimer's and Dementia, 2020, 16, e042104.	0.8	0