Tania F Gendron

List of Publications by Year
in descending order

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78 papers	10,747 citations	44069 48 h-index	69250 77 g-index
84 all docs	84 docs citations	84 times ranked	8071 citing authors

#	Article	IF	Citations
1	Unconventional Translation of C9ORF72 GGGGCC Expansion Generates Insoluble Polypeptides Specific to c9FTD/ALS. Neuron, 2013, 77, 639-646.	8.1	962
2	Targeting RNA Foci in iPSC-Derived Motor Neurons from ALS Patients with a C9ORF72 Repeat Expansion. Science Translational Medicine, 2013, 5, 208ra149.	12.4	586
3	Aberrant cleavage of TDP-43 enhances aggregation and cellular toxicity. Proceedings of the National Academy of Sciences of the United States of America, 2009, 106, 7607-7612.	7.1	523
4	Antisense transcripts of the expanded C9ORF72 hexanucleotide repeat form nuclear RNA foci and undergo repeat-associated non-ATG translation in c9FTD/ALS. Acta Neuropathologica, 2013, 126, 829-844.	7.7	506
5	Gain of Toxicity from ALS/FTD-Linked Repeat Expansions in C9ORF72 Is Alleviated by Antisense Oligonucleotides Targeting GGGGCC-Containing RNAs. Neuron, 2016, 90, 535-550.	8.1	437
6	The role of tau in neurodegeneration. Molecular Neurodegeneration, 2009, 4, 13.	10.8	353
7	C9ORF72 repeat expansions in mice cause TDP-43 pathology, neuronal loss, and behavioral deficits. Science, 2015, 348, 1151-1154.	12.6	332
8	Distinct brain transcriptome profiles in C9orf72-associated and sporadic ALS. Nature Neuroscience, 2015, 18, 1175-1182.	14.8	330
9	Poly(GR) in C9ORF72 -Related ALS/FTD Compromises Mitochondrial Function and Increases Oxidative Stress and DNA Damage in iPSC-Derived Motor Neurons. Neuron, 2016, 92, 383-391.	8.1	323
10	Discovery of a Biomarker and Lead Small Molecules to Target r(GGGGCC)-Associated Defects in c9FTD/ALS. Neuron, 2014, 83, 1043-1050.	8.1	289
11	Aggregation-prone c9FTD/ALS poly(GA) RAN-translated proteins cause neurotoxicity by inducing ER stress. Acta Neuropathologica, 2014, 128, 505-524.	7.7	284
12	C9ORF72 poly(GA) aggregates sequester and impair HR23 and nucleocytoplasmic transport proteins. Nature Neuroscience, 2016, 19, 668-677.	14.8	268
13	Reduced C9orf72 gene expression in c9FTD/ALS is caused by histone trimethylation, an epigenetic event detectable in blood. Acta Neuropathologica, 2013, 126, 895-905.	7.7	263
14	C9orf72 BAC Transgenic Mice Display Typical Pathologic Features of ALS/FTD. Neuron, 2015, 88, 892-901.	8.1	249
15	Poly(GR) impairs protein translation and stress granule dynamics in C9orf72-associated frontotemporal dementia and amyotrophic lateral sclerosis. Nature Medicine, 2018, 24, 1136-1142.	30.7	241
16	Amyotrophic Lateral Sclerosis: An Update for 2018. Mayo Clinic Proceedings, 2018, 93, 1617-1628.	3.0	227
17	Human C9ORF72 Hexanucleotide Expansion Reproduces RNA Foci and Dipeptide Repeat Proteins but Not Neurodegeneration in BAC Transgenic Mice. Neuron, 2015, 88, 902-909.	8.1	219
18	Quantitative analysis and clinico-pathological correlations of different dipeptide repeat protein pathologies in C9ORF72 mutation carriers. Acta Neuropathologica, 2015, 130, 845-861.	7.7	204

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19	Heterochromatin anomalies and double-stranded RNA accumulation underlie C9orf72 poly(PR) toxicity. Science, 2019, 363, .	12.6	181
20	Poly(GP) proteins are a useful pharmacodynamic marker for C9ORF72 -associated amyotrophic lateral sclerosis. Science Translational Medicine, 2017, 9, .	12.4	179
21	Differential Toxicity of Nuclear RNA Foci versus Dipeptide Repeat Proteins in a Drosophila Model of C9ORF72 FTD/ALS. Neuron, 2015, 87, 1207-1214.	8.1	176
22	Dipeptide repeat proteins are present in the p62 positive inclusions in patients with frontotemporal lobar degeneration and motor neurone disease associated with expansions in C9ORF72. Acta Neuropathologica Communications, 2013, 1, 68.	5.2	162
23	Reduced C9ORF72 function exacerbates gain of toxicity from ALS/FTD-causing repeat expansion in C9orf72. Nature Neuroscience, 2020, 23, 615-624.	14.8	157
24	The dual functions of the extreme N-terminus of TDP-43 in regulating its biological activity and inclusion formation. Human Molecular Genetics, 2013, 22, 3112-3122.	2.9	156
25	Timing and significance of pathological features inC9orf72expansion-associated frontotemporal dementia. Brain, 2016, 139, 3202-3216.	7.6	136
26	Mechanisms of toxicity in C9FTLD/ALS. Acta Neuropathologica, 2014, 127, 359-376.	7.7	134
27	CUG initiation and frameshifting enable production of dipeptide repeat proteins from ALS/FTD C9ORF72 transcripts. Nature Communications, 2018, 9, 152.	12.8	123
28	Truncated stathmin-2 is a marker of TDP-43 pathology in frontotemporal dementia. Journal of Clinical Investigation, 2020, 130, 6080-6092.	8.2	117
29	Spt4 selectively regulates the expression of C9orf72 sense and antisense mutant transcripts. Science, 2016, 353, 708-712.	12.6	116
30	C9orf72 poly(GR) aggregation induces TDP-43 proteinopathy. Science Translational Medicine, 2020, 12, .	12.4	115
31	Long-read sequencing across the C9orf72 â€˜GGGGCCâ€™ repeat expansion: implications for clinical use and genetic discovery efforts in human disease. Molecular Neurodegeneration, 2018, 13, 46.	10.8	111
32	Aberrant deposition of stress granule-resident proteins linked to C9orf72-associated TDP-43 proteinopathy. Molecular Neurodegeneration, 2019, 14, 9.	10.8	111
33	Novel clinical associations with specific C9ORF72 transcripts in patients with repeat expansions in C9ORF72. Acta Neuropathologica, 2015, 130, 863-876.	7.7	104
34	Repetitive element transcripts are elevated in the brain of C9orf72 ALS/FTLD patients. Human Molecular Genetics, 2017, 26, 3421-3431.	2.9	101
35	Spinal poly-GA inclusions in a C9orf72 mouse model trigger motor deficits and inflammation without neuron loss. Acta Neuropathologica, 2017, 134, 241-254.	7.7	99
36	A zebrafish model for C9orf72 ALS reveals RNA toxicity as a pathogenic mechanism. Acta Neuropathologica, 2018, 135, 427-443.	7.7	98

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37	Cerebellar c9RAN proteins associate with clinical and neuropathological characteristics of C9ORF72 repeat expansion carriers. Acta Neuropathologica, 2015, 130, 559-573.	7.7	89
38	Phosphorylated neurofilament heavy chain: A biomarker of survival for <scp>C9ORF</scp>72â€associated amyotrophic lateral sclerosis. Annals of Neurology, 2017, 82, 139-146.	5.3	88
39	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
40	Misregulation of human sortilin splicing leads to the generation of a nonfunctional progranulin receptor. Proceedings of the National Academy of Sciences of the United States of America, 2012, 109, 21510-21515.	7.1	82
41	The Hairpin Form of r(G4C2)exp in c9ALS/FTD Is Repeat-Associated Non-ATG Translated and a Target for Bioactive Small Molecules. Cell Chemical Biology, 2019, 26, 179-190.e12.	5.2	80
42	Poly-GR dipeptide repeat polymers correlate with neurodegeneration and Clinicopathological subtypes in C9ORF72-related brain disease. Acta Neuropathologica Communications, 2018, 6, 63.	5.2	79
43	In-depth clinico-pathological examination of RNA foci in a large cohort of C9ORF72 expansion carriers. Acta Neuropathologica, 2017, 134, 255-269.	7.7	76
44	Disease Mechanisms ofC9ORF72Repeat Expansions. Cold Spring Harbor Perspectives in Medicine, 2018, 8, a024224.	6.2	75
45	Characterization of DNA hypermethylation in the cerebellum of c9FTD/ALS patients. Brain Research, 2014, 1584, 15-21.	2.2	70
46	Serum neurofilament light protein correlates with unfavorable clinical outcomes in hospitalized patients with COVID-19. Science Translational Medicine, 2021, 13, .	12.4	67
47	<scp>TDP</scp>â€1, the <scp>C</scp>aenorhabditis elegans ortholog of <scp>TDP</scp>â€43, limits the accumulation of doubleâ€stranded <scp>RNA</scp>. EMBO Journal, 2014, 33, 2947-2966.	7.8	62
48	Rodent Models of TDP-43 Proteinopathy: Investigating the Mechanisms of TDP-43-Mediated Neurodegeneration. Journal of Molecular Neuroscience, 2011, 45, 486-499.	2.3	59
49	Dipeptide repeat proteins inhibit homology-directed DNA double strand break repair in C9ORF72 ALS/FTD. Molecular Neurodegeneration, 2020, 15, 13.	10.8	58
50	Nucleocytoplasmic Proteomic Analysis Uncovers eRF1 and Nonsense-Mediated Decay as Modifiers of ALS/FTD C9orf72 Toxicity. Neuron, 2020, 106, 90-107.e13.	8.1	58
51	Cross-sectional and longitudinal measures of chitinase proteins in amyotrophic lateral sclerosis and expression of CHI3L1 in activated astrocytes. Journal of Neurology, Neurosurgery and Psychiatry, 2020, 91, 350-358.	1.9	54
52	Plasma neurofilament light predicts mortality in patients with stroke. Science Translational Medicine, 2020, 12, .	12.4	51
53	Chimeric Peptide Species Contribute to Divergent Dipeptide Repeat Pathology in c9ALS/FTD and SCA36. Neuron, 2020, 107, 292-305.e6.	8.1	51
54	Poly(GP), neurofilament and grey matter deficits in C9orf72 expansion carriers. Annals of Clinical and Translational Neurology, 2018, 5, 583-597.	3.7	48

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55	The extreme N-terminus of TDP-43 mediates the cytoplasmic aggregation of TDP-43 and associated toxicity in vivo. Brain Research, 2016, 1647, 57-64.	2.2	44
56	TARDBP Mutation Analysis in TDP-43 Proteinopathies and Deciphering the Toxicity of Mutant TDP-43. Journal of Alzheimer's Disease, 2012, 33, S35-S45.	2.6	43
57	Extensive transcriptomic study emphasizes importance of vesicular transport in C9orf72 expansion carriers. Acta Neuropathologica Communications, 2019, 7, 150.	5.2	40
58	Transcription elongation factor AFF2/FMR2 regulates expression of expanded GGGGCC repeat-containing C9ORF72 allele in ALS/FTD. Nature Communications, 2019, 10, 5466.	12.8	40
59	Ribonuclease recruitment using a small molecule reduced c9ALS/FTD r(G ₄ C ₂) Tj ETQq1 1 0.784314 rgBT /O	12.4	39
60	Hexanucleotide Repeat Expansions in c9FTD/ALS and SCA36 Confer Selective Patterns of Neurodegeneration InÂVivo. Cell Reports, 2020, 31, 107616.	6.4	37
61	Toward allele-specific targeting therapy and pharmacodynamic marker for spinocerebellar ataxia type 3. Science Translational Medicine, 2020, 12, .	12.4	32
62	Sensitive ELISA-based detection method for the mitophagy marker p-S65-Ub in human cells, autopsy brain, and blood samples. Autophagy, 2021, 17, 2613-2628.	9.1	29
63	Biomarkers for Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Associated With Hexanucleotide Expansion Mutations in C9orf72. Frontiers in Neurology, 2018, 9, 1063.	2.4	28
64	A C. elegans model of C9orf72-associated ALS/FTD uncovers a conserved role for eIF2D in RAN translation. Nature Communications, 2021, 12, 6025.	12.8	27
65	A C9ORF72 BAC mouse model recapitulates key epigenetic perturbations of ALS/FTD. Molecular Neurodegeneration, 2017, 12, 46.	10.8	22
66	Ethanol enhances tau accumulation in neuroblastoma cells that inducibly express tau. Neuroscience Letters, 2008, 443, 67-71.	2.1	21
67	Comprehensive cross-sectional and longitudinal analyses of plasma neurofilament light across FTD spectrum disorders. Cell Reports Medicine, 2022, 3, 100607.	6.5	21
68	Loss of Tmem106b is unable to ameliorate frontotemporal dementia-like phenotypes in an AAV mouse model of C9ORF72-repeat induced toxicity. Acta Neuropathologica Communications, 2018, 6, 42.	5.2	20
69	Deep vein thrombosis and pulmonary embolism among hospitalized coronavirus disease 2019â€“positive patients predicted for higher mortality and prolonged intensive care unit and hospital stays in a multisite healthcare system. Journal of Vascular Surgery: Venous and Lymphatic Disorders, 2021, 9, 1361-1370.e1.	1.6	17
70	Long-read targeted sequencing uncovers clinicopathological associations for C9orf72-linked diseases. Brain, 2021, 144, 1082-1088.	7.6	17
71	Abnormal expression of homeobox genes and transthyretin in C9ORF72 expansion carriers. Neurology: Genetics, 2017, 3, e161.	1.9	12
72	OPTN p.Met468Arg and ATXN2 intermediate length polyQ extension in families with C9orf72 mediated amyotrophic lateral sclerosis and frontotemporal dementia. American Journal of Medical Genetics Part B: Neuropsychiatric Genetics, 2018, 177, 75-85.	1.7	12

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73	Structural Features of Small Molecules Targeting the RNA Repeat Expansion That Causes Genetically Defined ALS/FTD. ACS Chemical Biology, 2020, 15, 3112-3123.	3.4	12
74	Alterations of mesenchymal stromal cells in cerebrospinal fluid: insights from transcriptomics and an ALS clinical trial. Stem Cell Research and Therapy, 2021, 12, 187.	5.5	8
75	A Small Molecule Exploits Hidden Structural Features within the RNA Repeat Expansion That Causes c9ALS/FTD and Rescues Pathological Hallmarks. ACS Chemical Neuroscience, 2021, 12, 4076-4089.	3.5	8
76	ARHGEF28 p.Lys280Metfs40Ter in an amyotrophic lateral sclerosis family with a C9orf72 expansion. Neurology: Genetics, 2017, 3, e190.	1.9	6
77	Does Obesity-Induced Â Phosphorylation Tip the Scale Toward Dementia?. Diabetes, 2013, 62, 1365-1366.	0.6	4
78	Cover Image, Volume 177B, Number 1, January 2018. American Journal of Medical Genetics Part B: Neuropsychiatric Genetics, 2018, 177, i.	1.7	0