

# Ian R A Mackenzie

## List of Publications by Year in descending order

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161  
papers

34,766  
citations

9264

74  
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6654

156  
g-index

171  
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171  
docs citations

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times ranked

20580  
citing authors

#	ARTICLE	IF	CITATIONS
1	Ubiquitinated TDP-43 in Frontotemporal Lobar Degeneration and Amyotrophic Lateral Sclerosis. <i>Science</i> , 2006, 314, 130-133.	12.6	5,422
2	Expanded GGGGCC Hexanucleotide Repeat in Noncoding Region of C9ORF72 Causes Chromosome 9p-Linked FTD and ALS. <i>Neuron</i> , 2011, 72, 245-256.	8.1	4,176
3	Mutations in progranulin cause tau-negative frontotemporal dementia linked to chromosome 17. <i>Nature</i> , 2006, 442, 916-919.	27.8	1,816
4	Correlation of Alzheimer Disease Neuropathologic Changes With Cognitive Status: A Review of the Literature. <i>Journal of Neuropathology and Experimental Neurology</i> , 2012, 71, 362-381.	1.7	1,599
5	Neuropathologic diagnostic and nosologic criteria for frontotemporal lobar degeneration: consensus of the Consortium for Frontotemporal Lobar Degeneration. <i>Acta Neuropathologica</i> , 2007, 114, 5-22.	7.7	978
6	Nomenclature and nosology for neuropathologic subtypes of frontotemporal lobar degeneration: an update. <i>Acta Neuropathologica</i> , 2010, 119, 1-4.	7.7	854
7	Pathological TDP-43 distinguishes sporadic amyotrophic lateral sclerosis from amyotrophic lateral sclerosis with <i>SOD1</i> mutations. <i>Annals of Neurology</i> , 2007, 61, 427-434.	5.3	840
8	A harmonized classification system for FTLD-TDP pathology. <i>Acta Neuropathologica</i> , 2011, 122, 111-113.	7.7	817
9	ALS-associated fused in sarcoma (FUS) mutations disrupt Transportin-mediated nuclear import. <i>EMBO Journal</i> , 2010, 29, 2841-2857.	7.8	717
10	A new subtype of frontotemporal lobar degeneration with FUS pathology. <i>Brain</i> , 2009, 132, 2922-2931.	7.6	628
11	Progranulin Deficiency Promotes Circuit-Specific Synaptic Pruning by Microglia via Complement Activation. <i>Cell</i> , 2016, 165, 921-935.	28.9	558
12	Mutations in progranulin are a major cause of ubiquitin-positive frontotemporal lobar degeneration. <i>Human Molecular Genetics</i> , 2006, 15, 2988-3001.	2.9	529
13	TIA1 Mutations in Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Promote Phase Separation and Alter Stress Granule Dynamics. <i>Neuron</i> , 2017, 95, 808-816.e9.	8.1	493
14	Common variants at 7p21 are associated with frontotemporal lobar degeneration with TDP-43 inclusions. <i>Nature Genetics</i> , 2010, 42, 234-239.	21.4	479
15	Sortilin-Mediated Endocytosis Determines Levels of the Frontotemporal Dementia Protein, Progranulin. <i>Neuron</i> , 2010, 68, 654-667.	8.1	465
16	TDP-43 in Familial and Sporadic Frontotemporal Lobar Degeneration with Ubiquitin Inclusions. <i>American Journal of Pathology</i> , 2007, 171, 227-240.	3.8	446
17	Novel Mutations in TARDBP (TDP-43) in Patients with Familial Amyotrophic Lateral Sclerosis. <i>PLoS Genetics</i> , 2008, 4, e1000193.	3.5	393
18	Ageing-related tau astrogliaopathy (ARTAG): harmonized evaluation strategy. <i>Acta Neuropathologica</i> , 2016, 131, 87-102.	7.7	380

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19	Nomenclature for neuropathologic subtypes of frontotemporal lobar degeneration: consensus recommendations. <i>Acta Neuropathologica</i> , 2009, 117, 15-18.	7.7	377
20	Neuropathological background of phenotypical variability in frontotemporal dementia. <i>Acta Neuropathologica</i> , 2011, 122, 137-153.	7.7	375
21	Advances in understanding the molecular basis of frontotemporal dementia. <i>Nature Reviews Neurology</i> , 2012, 8, 423-434.	10.1	353
22	hnRNP A3 binds to GGGGCC repeats and is a constituent of p62-positive/TDP43-negative inclusions in the hippocampus of patients with C9orf72 mutations. <i>Acta Neuropathologica</i> , 2013, 125, 413-423.	7.7	302
23	Frontotemporal dementia and its subtypes: a genome-wide association study. <i>Lancet Neurology</i> , The, 2014, 13, 686-699.	10.2	302
24	Heterogeneity of ubiquitin pathology in frontotemporal lobar degeneration: classification and relation to clinical phenotype. <i>Acta Neuropathologica</i> , 2006, 112, 539-549.	7.7	298
25	Dipeptide repeat protein pathology in C9ORF72 mutation cases: clinico-pathological correlations. <i>Acta Neuropathologica</i> , 2013, 126, 859-879.	7.7	298
26	TDP-43 in the Ubiquitin Pathology of Frontotemporal Dementia With VCP Gene Mutations. <i>Journal of Neuropathology and Experimental Neurology</i> , 2007, 66, 152-157.	1.7	295
27	Phosphorylated TDP-43 in Alzheimer's disease and dementia with Lewy bodies. <i>Acta Neuropathologica</i> , 2009, 117, 125-136.	7.7	294
28	The neuropathology of frontotemporal lobar degeneration caused by mutations in the progranulin gene. <i>Brain</i> , 2006, 129, 3081-3090.	7.6	291
29	Ubiquitinated pathological lesions in frontotemporal lobar degeneration contain the TAR DNA-binding protein, TDP-43. <i>Acta Neuropathologica</i> , 2007, 113, 521-533.	7.7	274
30	Common variation in the miR-659 binding-site of GRN is a major risk factor for TDP43-positive frontotemporal dementia. <i>Human Molecular Genetics</i> , 2008, 17, 3631-3642.	2.9	271
31	Arginine methylation next to the PY-NLS modulates Transportin binding and nuclear import of FUS. <i>EMBO Journal</i> , 2012, 31, 4258-4275.	7.8	266
32	Molecular neuropathology of frontotemporal dementia: insights into disease mechanisms from postmortem studies. <i>Journal of Neurochemistry</i> , 2016, 138, 54-70.	3.9	252
33	FET proteins TAF15 and EWS are selective markers that distinguish FTLD with FUS pathology from amyotrophic lateral sclerosis with FUS mutations. <i>Brain</i> , 2011, 134, 2595-2609.	7.6	247
34	Abundant FUS-immunoreactive pathology in neuronal intermediate filament inclusion disease. <i>Acta Neuropathologica</i> , 2009, 118, 605-616.	7.7	237
35	FUS pathology in basophilic inclusion body disease. <i>Acta Neuropathologica</i> , 2009, 118, 617-627.	7.7	222
36	FUS pathology defines the majority of tau- and TDP-43-negative frontotemporal lobar degeneration. <i>Acta Neuropathologica</i> , 2010, 120, 33-41.	7.7	222

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37	Novel Ubiquitin Neuropathology in Frontotemporal Dementia With Valosin-Containing Protein Gene Mutations. <i>Journal of Neuropathology and Experimental Neurology</i> , 2006, 65, 571-581.	1.7	206
38	Quantitative analysis and clinico-pathological correlations of different dipeptide repeat protein pathologies in C9ORF72 mutation carriers. <i>Acta Neuropathologica</i> , 2015, 130, 845-861.	7.7	204
39	Clinical and pathological features of familial frontotemporal dementia caused by C9ORF72 mutation on chromosome 9p. <i>Brain</i> , 2012, 135, 709-722.	7.6	201
40	Phenotypic variability associated with progranulin haploinsufficiency in patients with the common 1477C>T (Arg493X) mutation: an international initiative. <i>Lancet Neurology</i> , The, 2007, 6, 857-868.	10.2	199
41	Progranulin in frontotemporal lobar degeneration and neuroinflammation. <i>Journal of Neuroinflammation</i> , 2007, 4, 7.	7.2	194
42	Phenotypic Variability of Gerstmann-Straussler-Scheinker Disease is Associated with Prion Protein Heterogeneity. <i>Journal of Neuropathology and Experimental Neurology</i> , 1998, 57, 979-988.	1.7	182
43	Age at symptom onset and death and disease duration in genetic frontotemporal dementia: an international retrospective cohort study. <i>Lancet Neurology</i> , The, 2020, 19, 145-156.	10.2	175
44	Clinical, neuroimaging and neuropathological features of a new chromosome 9p-linked FTD-ALS family. <i>Journal of Neurology, Neurosurgery and Psychiatry</i> , 2011, 82, 196-203.	1.9	170
45	The neuropathology associated with repeat expansions in the C9ORF72 gene. <i>Acta Neuropathologica</i> , 2014, 127, 347-357.	7.7	164
46	Pathological heterogeneity in amyotrophic lateral sclerosis with FUS mutations: two distinct patterns correlating with disease severity and mutation. <i>Acta Neuropathologica</i> , 2011, 122, 87-98.	7.7	153
47	The role of transactive response DNA-binding protein-43 in amyotrophic lateral sclerosis and frontotemporal dementia. <i>Current Opinion in Neurology</i> , 2008, 21, 693-700.	3.6	150
48	Individuals with progranulin haploinsufficiency exhibit features of neuronal ceroid lipofuscinosis. <i>Science Translational Medicine</i> , 2017, 9, .	12.4	147
49	Synaptic dysfunction in progranulin-deficient mice. <i>Neurobiology of Disease</i> , 2012, 45, 711-722.	4.4	144
50	Ubiquitin Immunohistochemistry Suggests Classic Motor Neuron Disease, Motor Neuron Disease With Dementia, and Frontotemporal Dementia of the Motor Neuron Disease Type Represent a Clinicopathologic Spectrum. <i>Journal of Neuropathology and Experimental Neurology</i> , 2005, 64, 730-739.	1.7	143
51	Distinct pathological subtypes of FTLD-FUS. <i>Acta Neuropathologica</i> , 2011, 121, 207-218.	7.7	139
52	Clinical and pathological features of amyotrophic lateral sclerosis caused by mutation in the C9ORF72 gene on chromosome 9p. <i>Acta Neuropathologica</i> , 2012, 123, 409-417.	7.7	137
53	TMEM106B protects C9ORF72 expansion carriers against frontotemporal dementia. <i>Acta Neuropathologica</i> , 2014, 127, 397-406.	7.7	133
54	Prevalence of amyloid $\beta$ pathology in distinct variants of primary progressive aphasia. <i>Annals of Neurology</i> , 2018, 84, 729-740.	5.3	132

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55	Atypical frontotemporal lobar degeneration with ubiquitin-positive, TDP-43-negative neuronal inclusions. <i>Brain</i> , 2008, 131, 1282-1293.	7.6	131
56	Genome-wide Screen Identifies rs646776 near Sortilin as a Regulator of Progranulin Levels in Human Plasma. <i>American Journal of Human Genetics</i> , 2010, 87, 890-897.	6.2	130
57	Central neurocytoma. , 1999, 85, 1606-1610.		122
58	The neuropathology and clinical phenotype of FTD with progranulin mutations. <i>Acta Neuropathologica</i> , 2007, 114, 49-54.	7.7	119
59	A Reassessment of the Neuropathology of Frontotemporal Dementia Linked to Chromosome 3. <i>Journal of Neuropathology and Experimental Neurology</i> , 2007, 66, 884-891.	1.7	118
60	Î±-Internexin Is Present in the Pathological Inclusions of Neuronal Intermediate Filament Inclusion Disease. <i>American Journal of Pathology</i> , 2004, 164, 2153-2161.	3.8	116
61	Progranulin: normal function and role in neurodegeneration. <i>Journal of Neurochemistry</i> , 2008, 104, 287-297.	3.9	114
62	Genetic and Clinical Features of Progranulin-Associated Frontotemporal Lobar Degeneration. <i>Archives of Neurology</i> , 2011, 68, 488.	4.5	108
63	A family with tau-negative frontotemporal dementia and neuronal intranuclear inclusions linked to chromosome 17. <i>Brain</i> , 2006, 129, 853-867.	7.6	102
64	<i>Fus</i> gene mutations in familial and sporadic amyotrophic lateral sclerosis. <i>Muscle and Nerve</i> , 2010, 42, 170-176.	2.2	101
65	Potential genetic modifiers of disease risk and age at onset in patients with frontotemporal lobar degeneration and GRN mutations: a genome-wide association study. <i>Lancet Neurology</i> , The, 2018, 17, 548-558.	10.2	97
66	CNS-derived extracellular vesicles from superoxide dismutase 1 (SOD1)G93A ALS mice originate from astrocytes and neurons and carry misfolded SOD1. <i>Journal of Biological Chemistry</i> , 2019, 294, 3744-3759.	3.4	97
67	Targeted manipulation of the sortilin-progranulin axis rescues progranulin haploinsufficiency. <i>Human Molecular Genetics</i> , 2014, 23, 1467-1478.	2.9	96
68	Frontotemporal lobar degeneration: current perspectives. <i>Neuropsychiatric Disease and Treatment</i> , 2014, 10, 297.	2.2	95
69	Mutations in progranulin explain atypical phenotypes with variants in MAPT. <i>Brain</i> , 2006, 129, 3124-3126.	7.6	91
70	Genome-wide analyses as part of the international FTLD-TDP whole-genome sequencing consortium reveals novel disease risk factors and increases support for immune dysfunction in FTLD. <i>Acta Neuropathologica</i> , 2019, 137, 879-899.	7.7	90
71	Mechanisms of disease in frontotemporal lobar degeneration: gain of function versus loss of function effects. <i>Acta Neuropathologica</i> , 2012, 124, 373-382.	7.7	89
72	<sc>TMEM</sc>106B p.T185S regulates <sc>TMEM</sc>106B protein levels: implications for frontotemporal dementia. <i>Journal of Neurochemistry</i> , 2013, 126, 781-791.	3.9	87

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73	Novel antibodies reveal presynaptic localization of C9orf72 protein and reduced protein levels in C9orf72 mutation carriers. <i>Acta Neuropathologica Communications</i> , 2018, 6, 72.	5.2	87
74	FUS-immunoreactive Intranuclear Inclusions in Neurodegenerative Disease. <i>Brain Pathology</i> , 2010, 20, 589-597.	4.1	84
75	C9ORF72 repeat expansions in cases with previously identified pathogenic mutations. <i>Neurology</i> , 2013, 81, 1332-1341.	1.1	84
76	Length of normal alleles of C9ORF72 GGGGCC repeat do not influence disease phenotype. <i>Neurobiology of Aging</i> , 2012, 33, 2950.e5-2950.e7.	3.1	83
77	Reappraisal of TDP-43 pathology in FTLD-U subtypes. <i>Acta Neuropathologica</i> , 2017, 134, 79-96.	7.7	83
78	Dementia lacking distinctive histology (DLDH) revisited. <i>Acta Neuropathologica</i> , 2006, 112, 551-559.	7.7	80
79	TDP-43-negative FTLD-U is a significant new clinico-pathological subtype of FTLD. <i>Acta Neuropathologica</i> , 2008, 116, 147-157.	7.7	77
80	rs5848 polymorphism and serum progranulin level. <i>Journal of the Neurological Sciences</i> , 2011, 300, 28-32.	0.6	77
81	The molecular genetics and neuropathology of frontotemporal lobar degeneration: recent developments. <i>Neurogenetics</i> , 2007, 8, 237-248.	1.4	76
82	Monomethylated and unmethylated FUS exhibit increased binding to Transportin and distinguish FTLD-FUS from ALS-FUS. <i>Acta Neuropathologica</i> , 2016, 131, 587-604.	7.7	76
83	Transportin 1 accumulates specifically with FET proteins but no other transportin cargos in FTLD-FUS and is absent in FUS inclusions in ALS with FUS mutations. <i>Acta Neuropathologica</i> , 2012, 124, 705-716.	7.7	74
84	Ataxin-2 as potential disease modifier in C9ORF72 expansion carriers. <i>Neurobiology of Aging</i> , 2014, 35, 2421.e13-2421.e17.	3.1	74
85	FET proteins in frontotemporal dementia and amyotrophic lateral sclerosis. <i>Brain Research</i> , 2012, 1462, 40-43.	2.2	71
86	Homotypic fibrillization of TMEM106B across diverse neurodegenerative diseases. <i>Cell</i> , 2022, 185, 1346-1355.e15.	28.9	70
87	The molecular basis of frontotemporal dementia. <i>Expert Reviews in Molecular Medicine</i> , 2009, 11, e23.	3.9	69
88	Early dipeptide repeat pathology in a frontotemporal dementia kindred with C9ORF72 mutation and intellectual disability. <i>Acta Neuropathologica</i> , 2014, 127, 451-458.	7.7	67
89	Genetic modifiers in carriers of repeat expansions in the C9ORF72 gene. <i>Molecular Neurodegeneration</i> , 2014, 9, 38.	10.8	63
90	Senile plaques do not progressively accumulate with normal aging. <i>Acta Neuropathologica</i> , 1994, 87, 520-525.	7.7	61

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91	Anterior brain glucose hypometabolism predates dementia in progranulin mutation carriers. <i>Neurology</i> , 2013, 81, 1322-1331.	1.1	60
92	Frontotemporal degeneration, the next therapeutic frontier: Molecules and animal models for frontotemporal degeneration drug development. <i>Alzheimer's and Dementia</i> , 2013, 9, 176-188.	0.8	58
93	Absence of FUS-immunoreactive pathology in frontotemporal dementia linked to chromosome 3 (FTD-3) caused by mutation in the CHMP2B gene. <i>Acta Neuropathologica</i> , 2009, 118, 719-720.	7.7	56
94	The Comprehensive Assessment of Neurodegeneration and Dementia: Canadian Cohort Study. <i>Canadian Journal of Neurological Sciences</i> , 2019, 46, 499-511.	0.5	56
95	The Neuropathology of FTD Associated With ALS. <i>Alzheimer Disease and Associated Disorders</i> , 2007, 21, S44-S49.	1.3	55
96	Interactions between ALS-linked FUS and nucleoporins are associated with defects in the nucleocytoplasmic transport pathway. <i>Nature Neuroscience</i> , 2021, 24, 1077-1088.	14.8	54
97	?-Internexin aggregates are abundant in neuronal intermediate filament inclusion disease (NIFID) but rare in other neurodegenerative diseases. <i>Acta Neuropathologica</i> , 2004, 108, 213-23.	7.7	50
98	Jump from Pre-mutation to Pathologic Expansion in C9orf72. <i>American Journal of Human Genetics</i> , 2015, 96, 962-970.	6.2	50
99	Neuropathological heterogeneity in frontotemporal lobar degeneration with TDP-43 proteinopathy: a quantitative study of 94 cases using principal components analysis. <i>Journal of Neural Transmission</i> , 2010, 117, 227-239.	2.8	48
100	The advantages of frontotemporal degeneration drug development (part 2 of frontotemporal) Tj ETQq0 0 0 rgBT /Overlock 10 Tf 50 38	0.8	48
101	eEF2K inhibition blocks A $\beta$ 242 neurotoxicity by promoting an NRF2 antioxidant response. <i>Acta Neuropathologica</i> , 2017, 133, 101-119.	7.7	48
102	Activity of translation regulator eukaryotic elongation factor-2 kinase is increased in Parkinson disease brain and its inhibition reduces alpha synuclein toxicity. <i>Acta Neuropathologica Communications</i> , 2018, 6, 54.	5.2	48
103	The relationship between extramotor ubiquitin-immunoreactive neuronal inclusions and dementia in motor neuron disease. <i>Acta Neuropathologica</i> , 2003, 105, 98-102.	7.7	47
104	Sex differences in the prevalence of genetic mutations in FTD and ALS. <i>Neurology</i> , 2017, 89, 1633-1642.	1.1	47
105	LATE to the PART-y. <i>Brain</i> , 2019, 142, e47-e47.	7.6	44
106	Two cases of rheumatoid meningitis. <i>Neuropathology</i> , 2016, 36, 93-102.	1.2	43
107	Amyloid Beta Immunoreactivity in the Retinal Ganglion Cell Layer of the Alzheimer's Eye. <i>Frontiers in Neuroscience</i> , 2020, 14, 758.	2.8	42
108	Reduced hnRNP A3 increases C9orf72 repeat RNA levels and dipeptide repeat protein deposition. <i>EMBO Reports</i> , 2016, 17, 1314-1325.	4.5	39

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109	Clinical and neuropathological features of ALS/FTD with TIA1 mutations. <i>Acta Neuropathologica Communications</i> , 2017, 5, 96.	5.2	38
110	Individualized atrophy scores predict dementia onset in familial frontotemporal lobar degeneration. <i>Alzheimer's and Dementia</i> , 2020, 16, 37-48.	0.8	38
111	Neuronal intranuclear inclusions distinguish familial FTD-MND type from sporadic cases. <i>Acta Neuropathologica</i> , 2003, 105, 543-548.	7.7	37
112	Subcortical TDP-43 pathology patterns validate cortical FTLD-TDP subtypes and demonstrate unique aspects of C9orf72 mutation cases. <i>Acta Neuropathologica</i> , 2020, 139, 83-98.	7.7	37
113	Development and validation of a novel dementia of Alzheimer's type (DAT) score based on metabolism FDG-PET imaging. <i>NeuroImage: Clinical</i> , 2018, 18, 802-813.	2.7	35
114	Novel Types of Frontotemporal Lobar Degeneration: Beyond Tau and TDP-43. <i>Journal of Molecular Neuroscience</i> , 2011, 45, 402-408.	2.3	33
115	Microglial lysosome dysfunction contributes to white matter pathology and TDP-43 proteinopathy in GRN-associated FTD. <i>Cell Reports</i> , 2021, 36, 109581.	6.4	33
116	Assessment of executive function declines in presymptomatic and mildly symptomatic familial frontotemporal dementia: NIHâ€œEXAMINER as a potential clinical trial endpoint. <i>Alzheimer's and Dementia</i> , 2020, 16, 11-21.	0.8	32
117	Spinal cord homogenates from SOD1 familial amyotrophic lateral sclerosis induce SOD1 aggregation in living cells. <i>PLoS ONE</i> , 2017, 12, e0184384.	2.5	32
118	Proposed research criteria for prodromal behavioural variant frontotemporal dementia. <i>Brain</i> , 2022, 145, 1079-1097.	7.6	30
119	Clinical and volumetric changes with increasing functional impairment in familial frontotemporal lobar degeneration. <i>Alzheimer's and Dementia</i> , 2020, 16, 49-59.	0.8	27
120	Gray matter changes in asymptomatic C9orf72 and GRN mutation carriers. <i>NeuroImage: Clinical</i> , 2018, 18, 591-598.	2.7	26
121	Frontotemporal Lobar Degeneration TDP-43-Immunoreactive Pathological Subtypes: Clinical and Mechanistic Significance. <i>Advances in Experimental Medicine and Biology</i> , 2021, 1281, 201-217.	1.6	26
122	Fused in Sarcoma Neuropathology in Neurodegenerative Disease. <i>Cold Spring Harbor Perspectives in Medicine</i> , 2017, 7, a024299.	6.2	25
123	Clinicopathologic correlations in a family with a <i>TBK1</i> mutation presenting as primary progressive aphasia and primary lateral sclerosis. <i>Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration</i> , 2019, 20, 568-575.	1.7	24
124	Drusen in the Peripheral Retina of the Alzheimerâ€™s Eye. <i>Current Alzheimer Research</i> , 2018, 15, 743-750.	1.4	24
125	Extrapyramidal features in patients with motor neuron disease and dementia; a clinicopathological correlative study. <i>Acta Neuropathologica</i> , 2004, 107, 336-340.	7.7	23
126	The spectrum and severity of FUS-immunoreactive inclusions in the frontal and temporal lobes of ten cases of neuronal intermediate filament inclusion disease. <i>Acta Neuropathologica</i> , 2011, 121, 219-228.	7.7	23



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127	Antibody against TDP-43 phosphorylated at serine 375 suggests conformational differences of TDP-43 aggregates among FTLDâ€“TDP subtypes. <i>Acta Neuropathologica</i> , 2020, 140, 645-658.	7.7	23
128	Neuronal Intranuclear Inclusions Distinguish Familial FTD-MND Type from Sporadic Cases. <i>Dementia and Geriatric Cognitive Disorders</i> , 2004, 17, 333-336.	1.5	22
129	Early Neuropsychological Characteristics of Progranulin Mutation Carriers. <i>Journal of the International Neuropsychological Society</i> , 2014, 20, 694-703.	1.8	21
130	Multiple brain pathologies in dementia are common. <i>European Geriatric Medicine</i> , 2010, 1, 259-265.	2.8	20
131	TDP-43 pathology in primary lateral sclerosis. <i>Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration</i> , 2020, 21, 52-58.	1.7	20
132	Rates of Brain Atrophy Across Disease Stages in Familial Frontotemporal Dementia Associated With MAPT, GRN, and C9orf72 Pathogenic Variants. <i>JAMA Network Open</i> , 2020, 3, e2022847.	5.9	19
133	Premature termination codon readthrough upregulates progranulin expression and improves lysosomal function in preclinical models of GRN deficiency. <i>Molecular Neurodegeneration</i> , 2020, 15, 21.	10.8	19
134	Progressive Anomia Revisited: Focal Degeneration Associated with Progranulin Gene Mutation. <i>Neurocase</i> , 2008, 13, 366-377.	0.6	17
135	Î±-Synuclein pathology in Parkinson disease activates homeostatic NRF2 anti-oxidant response. <i>Acta Neuropathologica Communications</i> , 2021, 9, 105.	5.2	17
136	FDG-PET in presymptomatic C9orf72 mutation carriers. <i>NeuroImage: Clinical</i> , 2021, 31, 102687.	2.7	16
137	Tracking white matter degeneration in asymptomatic and symptomatic MAPT mutation carriers. <i>Neurobiology of Aging</i> , 2019, 83, 54-62.	3.1	14
138	Mutations in protein N-arginine methyltransferases are not the cause of FTLD-FUS. <i>Neurobiology of Aging</i> , 2013, 34, 2235.e11-2235.e13.	3.1	13
139	Recognition memory and divergent cognitive profiles in prodromal genetic frontotemporal dementia. <i>Cortex</i> , 2021, 139, 99-115.	2.4	12
140	Developmental Delay, Treatment-Resistant Psychosis, and Early-Onset Dementia in a Man With 22q11 Deletion Syndrome and Huntingtonâ€™s Disease. <i>American Journal of Psychiatry</i> , 2018, 175, 400-407.	7.2	9
141	The contribution of behavioral features to caregiver burden in FTLD spectrum disorders. <i>Alzheimer's and Dementia</i> , 2022, 18, 1635-1649.	0.8	9
142	Neuropathology of primary lateral sclerosis. <i>Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration</i> , 2020, 21, 47-51.	1.7	8
143	The spectrum of disease and tau pathology of nodding syndrome in Uganda. <i>Brain</i> , 2023, 146, 954-967.	7.6	8
144	Spatial patterns of FUS-immunoreactive neuronal cytoplasmic inclusions (NCI) in neuronal intermediate filament inclusion disease (NIFID). <i>Journal of Neural Transmission</i> , 2011, 118, 1651-1657.	2.8	7

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145	Detection and characterization of TDP-43 in human cells and tissues by multiple reaction monitoring mass spectrometry. <i>Clinical Mass Spectrometry</i> , 2019, 14, 66-73.	1.9	7
146	Prodromal neuroinvasion of pathological $\alpha$ -synuclein in brainstem reticular nuclei and white matter lesions in a model of $\alpha$ -synucleinopathy. <i>Brain Communications</i> , 2021, 3, fcab104.	3.3	7
147	Familial frontotemporal dementia with neuronal intranuclear inclusions is not a polyglutamine expansion disease. <i>BMC Neurology</i> , 2006, 6, 32.	1.8	6
148	Decreased Prefrontal Activation during Matrix Reasoning in Predementia Progranulin Mutation Carriers. <i>Journal of Alzheimer's Disease</i> , 2018, 62, 583-589.	2.6	5
149	Expanding the Phenotype of Frontotemporal Lobar Degeneration With FUS-Positive Pathology (FTLD-FUS). <i>Journal of Neuropathology and Experimental Neurology</i> , 2020, 79, 809-812.	1.7	5
150	Aptamer-based enrichment of TDP-43 from human cells and tissues with quantification by HPLC-MS/MS. <i>Journal of Neuroscience Methods</i> , 2021, 363, 109344.	2.5	5
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