Ian R A Mackenzie

List of Publications by Year in Descending Order

161 papers	27,658 citations	71 h-index	166 g-index
171 ext. papers	31,527 ext. citations	9.8 avg, IF	6.42 L-index

#	Paper	IF	Citations
161	Proposed research criteria for prodromal behavioural variant frontotemporal dementia.. *Brain*, **2022**,	11.2	1
160	Chronic Traumatic Encephalopathy **2022**, 223-236		
159	Homotypic fibrillization of TMEM106B across diverse neurodegenerative diseases.. *Cell*, **2022**,	56.2	5
158	Longstanding Multiple System Atrophy-Parkinsonism with Limbic and FTLD-type Synuclein Pathology. *Neuropathology and Applied Neurobiology*, **2021**,	5.2	2
157	Left-Handed Man with Memory Complaints **2021**, 49-53		
156	Prodromal neuroinvasion of pathological synuclein in brainstem reticular nuclei and white matter lesions in a model of synucleinopathy. *Brain Communications*, **2021**, 3, fcab104	4.5	3
155	Interactions between ALS-linked FUS and nucleoporins are associated with defects in the nucleocytoplasmic transport pathway. *Nature Neuroscience*, **2021**, 24, 1077-1088	25.5	12
154	Synuclein pathology in Parkinson disease activates homeostatic NRF2 anti-oxidant response. *Acta Neuropathologica Communications*, **2021**, 9, 105	7.3	7
153	Recognition memory and divergent cognitive profiles in prodromal genetic frontotemporal dementia. *Cortex*, **2021**, 139, 99-115	3.8	3
152	Frontotemporal Lobar Degeneration TDP-43-Immunoreactive Pathological Subtypes: Clinical and Mechanistic Significance. *Advances in Experimental Medicine and Biology*, **2021**, 1281, 201-217	3.6	6
151	Microglial lysosome dysfunction contributes to white matter pathology and TDP-43 proteinopathy in GRN-associated FTD. *Cell Reports*, **2021**, 36, 109581	10.6	5
150	Aptamer-based enrichment of TDP-43 from human cells and tissues with quantification by HPLC-MS/MS. *Journal of Neuroscience Methods*, **2021**, 363, 109344	3	2
149	FDG-PET in presymptomatic C9orf72 mutation carriers. *NeuroImage: Clinical*, **2021**, 31, 102687	5.3	2
148	Rates of Brain Atrophy Across Disease Stages in Familial Frontotemporal Dementia Associated With MAPT, GRN, and C9orf72 Pathogenic Variants. *JAMA Network Open*, **2020**, 3, e2022847	10.4	5
147	Expanding the Phenotype of Frontotemporal Lobar Degeneration With FUS-Positive Pathology (FTLD-FUS). *Journal of Neuropathology and Experimental Neurology*, **2020**, 79, 809-812	3.1	3
146	Premature termination codon readthrough upregulates progranulin expression and improves lysosomal function in preclinical models of GRN deficiency. *Molecular Neurodegeneration*, **2020**, 15, 21	19	8
145	Applying the Alzheimer Disease ATN Diagnostic Framework in Atypical Dementia. *Alzheimer Disease and Associated Disorders*, **2020**, 34, 357-359	2.5	1

144	Neuropathology of primary lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2020**, 21, 47-51	3.6	2
143	Age at symptom onset and death and disease duration in genetic frontotemporal dementia: an international retrospective cohort study. *Lancet Neurology, The*, **2020**, 19, 145-156	24.1	90
142	Clinical and volumetric changes with increasing functional impairment in familial frontotemporal lobar degeneration. *Alzheimer's and Dementia*, **2020**, 16, 49-59	1.2	17
141	TDP-43 pathology in primary lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2020**, 21, 52-58	3.6	11
140	Antibody against TDP-43 phosphorylated at serine 375 suggests conformational differences of TDP-43 aggregates among FTLD-TDP subtypes. *Acta Neuropathologica*, **2020**, 140, 645-658	14.3	10
139	Amyloid Beta Immunoreactivity in the Retinal Ganglion Cell Layer of the Alzheimer's Eye. *Frontiers in Neuroscience*, **2020**, 14, 758	5.1	18
138	Assessment of executive function declines in presymptomatic and mildly symptomatic familial frontotemporal dementia: NIH-EXAMINER as a potential clinical trial endpoint. *Alzheimer's and Dementia*, **2020**, 16, 11-21	1.2	18
137	Individualized atrophy scores predict dementia onset in familial frontotemporal lobar degeneration. *Alzheimer's and Dementia*, **2020**, 16, 37-48	1.2	18
136	Subcortical TDP-43 pathology patterns validate cortical FTLD-TDP subtypes and demonstrate unique aspects of C9orf72 mutation cases. *Acta Neuropathologica*, **2020**, 139, 83-98	14.3	18
135	Tracking white matter degeneration in asymptomatic and symptomatic MAPT mutation carriers. *Neurobiology of Aging*, **2019**, 83, 54-62	5.6	9
134	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. *Acta Neuropathologica*, **2019**, 137, 879-899	14.3	50
133	A mitochondrial DNA D loop insertion detected almost exclusively in non-replicating tissues with maternal inheritance across three generations. *Mitochondrion*, **2019**, 46, 298-301	4.9	1
132	LATE to the PART-y. *Brain*, **2019**, 142, e47	11.2	25
131	The Comprehensive Assessment of Neurodegeneration and Dementia: Canadian Cohort Study. *Canadian Journal of Neurological Sciences*, **2019**, 46, 499-511	1	28
130	Detection and characterization of TDP-43 in human cells and tissues by multiple reaction monitoring mass spectrometry.. *Clinical Mass Spectrometry*, **2019**, 14 Pt B, 66-73	1.9	3
129	Clinicopathologic correlations in a family with a mutation presenting as primary progressive aphasia and primary lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2019**, 20, 568-575	3.6	16
128	Grant Report on PREDICT-ADFTD: Multimodal Imaging Prediction of AD/FTD and Differential Diagnosis. *Journal of Psychiatry and Brain Science*, **2019**, 4,	1.7	3
127	CNS-derived extracellular vesicles from superoxide dismutase 1 (SOD1) ALS mice originate from astrocytes and neurons and carry misfolded SOD1. *Journal of Biological Chemistry*, **2019**, 294, 3744-3759	5.4	59

126	Decreased Prefrontal Activation during Matrix Reasoning in Predementia Progranulin Mutation Carriers. *Journal of Alzheimer's Disease*, **2018**, 62, 583-589	4.3	3
125	Developmental Delay, Treatment-Resistant Psychosis, and Early-Onset Dementia in a Man With 22q11 Deletion Syndrome and Huntington's Disease. *American Journal of Psychiatry*, **2018**, 175, 400-407	11.9	7
124	Potential genetic modifiers of disease risk and age at onset in patients with frontotemporal lobar degeneration and GRN mutations: a genome-wide association study. *Lancet Neurology, The*, **2018**, 17, 548-558	24.1	60
123	Novel antibodies reveal presynaptic localization of C9orf72 protein and reduced protein levels in C9orf72 mutation carriers. *Acta Neuropathologica Communications*, **2018**, 6, 72	7.3	60
122	Activity of translation regulator eukaryotic elongation factor-2 kinase is increased in Parkinson disease brain and its inhibition reduces alpha synuclein toxicity. *Acta Neuropathologica Communications*, **2018**, 6, 54	7.3	27
121	Development and validation of a novel dementia of Alzheimer's type (DAT) score based on metabolism FDG-PET imaging. *NeuroImage: Clinical*, **2018**, 18, 802-813	5.3	22
120	Drusen in the Peripheral Retina of the Alzheimer's Eye. *Current Alzheimer Research*, **2018**, 15, 743-750	3	13
119	Prevalence of amyloid-β pathology in distinct variants of primary progressive aphasia. *Annals of Neurology*, **2018**, 84, 729-740	9.4	74
118	Gray matter changes in asymptomatic and mutation carriers. *NeuroImage: Clinical*, **2018**, 18, 591-598	5.3	16
117	Fused in Sarcoma Neuropathology in Neurodegenerative Disease. *Cold Spring Harbor Perspectives in Medicine*, **2017**, 7,	5.4	10
116	Individuals with progranulin haploinsufficiency exhibit features of neuronal ceroid lipofuscinosis. *Science Translational Medicine*, **2017**, 9,	17.5	107
115	Reappraisal of TDP-43 pathology in FTLD-U subtypes. *Acta Neuropathologica*, **2017**, 134, 79-96	14.3	66
114	Clinical and neuropathological features of ALS/FTD with TIA1 mutations. *Acta Neuropathologica Communications*, **2017**, 5, 96	7.3	27
113	Sex differences in the prevalence of genetic mutations in FTD and ALS: A meta-analysis. *Neurology*, **2017**, 89, 1633-1642	6.5	29
112	TIA1 Mutations in Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Promote Phase Separation and Alter Stress Granule Dynamics. *Neuron*, **2017**, 95, 808-816.e9	13.9	341
111	eEF2K inhibition blocks Aβ42 neurotoxicity by promoting an NRF2 antioxidant response. *Acta Neuropathologica*, **2017**, 133, 101-119	14.3	30
110	Spinal cord homogenates from SOD1 familial amyotrophic lateral sclerosis induce SOD1 aggregation in living cells. *PLoS ONE*, **2017**, 12, e0184384	3.7	22
109	Aging-related tau astrogliopathy (ARTAG): harmonized evaluation strategy. *Acta Neuropathologica*, **2016**, 131, 87-102	14.3	272

108	Monomethylated and unmethylated FUS exhibit increased binding to Transportin and distinguish FTLD-FUS from ALS-FUS. *Acta Neuropathologica*, **2016**, 131, 587-604	14.3	56
107	Molecular neuropathology of frontotemporal dementia: insights into disease mechanisms from postmortem studies. *Journal of Neurochemistry*, **2016**, 138 Suppl 1, 54-70	6	167
106	An Unusual Case of Rabies Encephalitis. *Canadian Journal of Neurological Sciences*, **2016**, 43, 852-855	1	2
105	Reduced hnRNPA3 increases C9orf72 repeat RNA levels and dipeptide-repeat protein deposition. *EMBO Reports*, **2016**, 17, 1314-25	6.5	29
104	Progranulin Deficiency Promotes Circuit-Specific Synaptic Pruning by Microglia via Complement Activation. *Cell*, **2016**, 165, 921-35	56.2	378
103	Two cases of rheumatoid meningitis. *Neuropathology*, **2016**, 36, 93-102	2	30
102	Quantitative analysis and clinico-pathological correlations of different dipeptide repeat protein pathologies in C9ORF72 mutation carriers. *Acta Neuropathologica*, **2015**, 130, 845-61	14.3	155
101	Jump from pre-mutation to pathologic expansion in C9orf72. *American Journal of Human Genetics*, **2015**, 96, 962-70	11	41
100	The neuropathology associated with repeat expansions in the C9ORF72 gene. *Acta Neuropathologica*, **2014**, 127, 347-57	14.3	135
99	Early dipeptide repeat pathology in a frontotemporal dementia kindred with C9ORF72 mutation and intellectual disability. *Acta Neuropathologica*, **2014**, 127, 451-8	14.3	56
98	TMEM106B protects C9ORF72 expansion carriers against frontotemporal dementia. *Acta Neuropathologica*, **2014**, 127, 397-406	14.3	108
97	Ataxin-2 as potential disease modifier in C9ORF72 expansion carriers. *Neurobiology of Aging*, **2014**, 35, 2421.e13-7	5.6	62
96	Frontotemporal dementia and its subtypes: a genome-wide association study. *Lancet Neurology, The*, **2014**, 13, 686-99	24.1	207
95	Genetic modifiers in carriers of repeat expansions in the C9ORF72 gene. *Molecular Neurodegeneration*, **2014**, 9, 38	19	51
94	Targeted manipulation of the sortilin-progranulin axis rescues progranulin haploinsufficiency. *Human Molecular Genetics*, **2014**, 23, 1467-78	5.6	71
93	Frontotemporal lobar degeneration: current perspectives. *Neuropsychiatric Disease and Treatment*, **2014**, 10, 297-310	3.1	79
92	Early neuropsychological characteristics of progranulin mutation carriers. *Journal of the International Neuropsychological Society*, **2014**, 20, 694-703	3.1	15
91	The advantages of frontotemporal degeneration drug development (part 2 of frontotemporal degeneration: the next therapeutic frontier). *Alzheimers and Dementia*, **2013**, 9, 189-98	1.2	42

90	Frontotemporal degeneration, the next therapeutic frontier: molecules and animal models for frontotemporal degeneration drug development. *Alzheimers and Dementia*, **2013**, 9, 176-88	1.2	45
89	Mutations in protein N-arginine methyltransferases are not the cause of FTLD-FUS. *Neurobiology of Aging*, **2013**, 34, 2235.e11-3	5.6	11
88	hnRNP A3 binds to GGGGCC repeats and is a constituent of p62-positive/TDP43-negative inclusions in the hippocampus of patients with C9orf72 mutations. *Acta Neuropathologica*, **2013**, 125, 413-23	14.3	250
87	Dipeptide repeat protein pathology in C9ORF72 mutation cases: clinico-pathological correlations. *Acta Neuropathologica*, **2013**, 126, 859-79	14.3	248
86	C9ORF72 repeat expansions in cases with previously identified pathogenic mutations. *Neurology*, **2013**, 81, 1332-41	6.5	75
85	Anterior brain glucose hypometabolism predates dementia in progranulin mutation carriers. *Neurology*, **2013**, 81, 1322-31	6.5	45
84	TMEM106B p.T185S regulates TMEM106B protein levels: implications for frontotemporal dementia. *Journal of Neurochemistry*, **2013**, 126, 781-91	6	57
83	FET proteins in frontotemporal dementia and amyotrophic lateral sclerosis. *Brain Research*, **2012**, 1462, 40-3	3.7	60
82	Synaptic dysfunction in progranulin-deficient mice. *Neurobiology of Disease*, **2012**, 45, 711-22	7.5	119
81	Clinical and pathological features of amyotrophic lateral sclerosis caused by mutation in the C9ORF72 gene on chromosome 9p. *Acta Neuropathologica*, **2012**, 123, 409-17	14.3	127
80	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. *Acta Neuropathologica*, **2012**, 124, 705-16	14.3	64
79	Mechanisms of disease in frontotemporal lobar degeneration: gain of function versus loss of function effects. *Acta Neuropathologica*, **2012**, 124, 373-82	14.3	69
78	Length of normal alleles of C9ORF72 GGGGCC repeat do not influence disease phenotype. *Neurobiology of Aging*, **2012**, 33, 2950.e5-7	5.6	72
77	Advances in understanding the molecular basis of frontotemporal dementia. *Nature Reviews Neurology*, **2012**, 8, 423-34	15	306
76	Arginine methylation next to the PY-NLS modulates Transportin binding and nuclear import of FUS. *EMBO Journal*, **2012**, 31, 4258-75	13	211
75	Clinical and pathological features of familial frontotemporal dementia caused by C9ORF72 mutation on chromosome 9p. *Brain*, **2012**, 135, 709-22	11.2	172
74	Correlation of Alzheimer disease neuropathologic changes with cognitive status: a review of the literature. *Journal of Neuropathology and Experimental Neurology*, **2012**, 71, 362-81	3.1	1145
73	rs5848 polymorphism and serum progranulin level. *Journal of the Neurological Sciences*, **2011**, 300, 28-32	3.2	64

72	Expanded GGGGCC hexanucleotide repeat in noncoding region of C9ORF72 causes chromosome 9p-linked FTD and ALS. *Neuron*, **2011**, 72, 245-56	13.9	3267
71	The spectrum and severity of FUS-immunoreactive inclusions in the frontal and temporal lobes of ten cases of neuronal intermediate filament inclusion disease. *Acta Neuropathologica*, **2011**, 121, 219-28	14.3	20
70	Distinct pathological subtypes of FTLD-FUS. *Acta Neuropathologica*, **2011**, 121, 207-18	14.3	116
69	Pathological heterogeneity in amyotrophic lateral sclerosis with FUS mutations: two distinct patterns correlating with disease severity and mutation. *Acta Neuropathologica*, **2011**, 122, 87-98	14.3	128
68	Neuropathological background of phenotypical variability in frontotemporal dementia. *Acta Neuropathologica*, **2011**, 122, 137-53	14.3	311
67	A harmonized classification system for FTLD-TDP pathology. *Acta Neuropathologica*, **2011**, 122, 111-3	14.3	656
66	Spatial patterns of FUS-immunoreactive neuronal cytoplasmic inclusions (NCI) in neuronal intermediate filament inclusion disease (NIFID). *Journal of Neural Transmission*, **2011**, 118, 1651-7	4.3	5
65	Novel types of frontotemporal lobar degeneration: beyond tau and TDP-43. *Journal of Molecular Neuroscience*, **2011**, 45, 402-8	3.3	30
64	Genetic and clinical features of progranulin-associated frontotemporal lobar degeneration. *Archives of Neurology*, **2011**, 68, 488-97		93
63	FET proteins TAF15 and EWS are selective markers that distinguish FTLD with FUS pathology from amyotrophic lateral sclerosis with FUS mutations. *Brain*, **2011**, 134, 2595-609	11.2	204
62	Clinical, neuroimaging and neuropathological features of a new chromosome 9p-linked FTD-ALS family. *Journal of Neurology, Neurosurgery and Psychiatry*, **2011**, 82, 196-203	5.5	146
61	ALS-associated fused in sarcoma (FUS) mutations disrupt Transportin-mediated nuclear import. *EMBO Journal*, **2010**, 29, 2841-57	13	587
60	Common variants at 7p21 are associated with frontotemporal lobar degeneration with TDP-43 inclusions. *Nature Genetics*, **2010**, 42, 234-9	36.3	361
59	FUS-immunoreactive intranuclear inclusions in neurodegenerative disease. *Brain Pathology*, **2010**, 20, 589-97	6	78
58	Multiple brain pathologies in dementia are common. *European Geriatric Medicine*, **2010**, 1, 259-265	3	17
57	Sortilin-mediated endocytosis determines levels of the frontotemporal dementia protein, progranulin. *Neuron*, **2010**, 68, 654-67	13.9	368
56	Genome-wide screen identifies rs646776 near sortilin as a regulator of progranulin levels in human plasma. *American Journal of Human Genetics*, **2010**, 87, 890-7	11	110
55	Nomenclature and nosology for neuropathologic subtypes of frontotemporal lobar degeneration: an update. *Acta Neuropathologica*, **2010**, 119, 1-4	14.3	711

54	FUS pathology defines the majority of tau- and TDP-43-negative frontotemporal lobar degeneration. *Acta Neuropathologica*, **2010**, 120, 33-41	14.3	198
53	Neuropathological heterogeneity in frontotemporal lobar degeneration with TDP-43 proteinopathy: a quantitative study of 94 cases using principal components analysis. *Journal of Neural Transmission*, **2010**, 117, 227-39	4.3	47
52	Fus gene mutations in familial and sporadic amyotrophic lateral sclerosis. *Muscle and Nerve*, **2010**, 42, 170-6	3.4	85
51	The molecular basis of frontotemporal dementia. *Expert Reviews in Molecular Medicine*, **2009**, 11, e23	6.7	57
50	A new subtype of frontotemporal lobar degeneration with FUS pathology. *Brain*, **2009**, 132, 2922-31	11.2	535
49	Nomenclature for neuropathologic subtypes of frontotemporal lobar degeneration: consensus recommendations. *Acta Neuropathologica*, **2009**, 117, 15-8	14.3	325
48	Phosphorylated TDP-43 in Alzheimer's disease and dementia with Lewy bodies. *Acta Neuropathologica*, **2009**, 117, 125-36	14.3	248
47	Abundant FUS-immunoreactive pathology in neuronal intermediate filament inclusion disease. *Acta Neuropathologica*, **2009**, 118, 605-16	14.3	213
46	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, 719-20	14.3	51
45	FUS pathology in basophilic inclusion body disease. *Acta Neuropathologica*, **2009**, 118, 617-27	14.3	198
44	Atypical frontotemporal lobar degeneration with ubiquitin-positive, TDP-43-negative neuronal inclusions. *Brain*, **2008**, 131, 1282-93	11.2	120
43	Novel mutations in TARDBP (TDP-43) in patients with familial amyotrophic lateral sclerosis. *PLoS Genetics*, **2008**, 4, e1000193	6	339
42	Common variation in the miR-659 binding-site of GRN is a major risk factor for TDP43-positive frontotemporal dementia. *Human Molecular Genetics*, **2008**, 17, 3631-42	5.6	242
41	The role of transactive response DNA-binding protein-43 in amyotrophic lateral sclerosis and frontotemporal dementia. *Current Opinion in Neurology*, **2008**, 21, 693-700	7.1	91
40	Progranulin: normal function and role in neurodegeneration. *Journal of Neurochemistry*, **2008**, 104, 287-97	4.7	96
39	TDP-43-negative FTLD-U is a significant new clinico-pathological subtype of FTLD. *Acta Neuropathologica*, **2008**, 116, 147-57	14.3	72
38	Progressive anomia revisited: focal degeneration associated with progranulin gene mutation. *Neurocase*, **2007**, 13, 366-77	0.8	12
37	Progranulin in frontotemporal lobar degeneration and neuroinflammation. *Journal of Neuroinflammation*, **2007**, 4, 7	10.1	172

36	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
35	Phenotypic variability associated with progranulin haploinsufficiency in patients with the common 1477C-->T (Arg493X) mutation: an international initiative. *Lancet Neurology, The*, **2007**, 6, 857-68	24.1	174
34	Ubiquitinated pathological lesions in frontotemporal lobar degeneration contain the TAR DNA-binding protein, TDP-43. *Acta Neuropathologica*, **2007**, 113, 521-33	14.3	252
33	The neuropathology and clinical phenotype of FTD with progranulin mutations. *Acta Neuropathologica*, **2007**, 114, 49-54	14.3	101
32	Neuropathologic diagnostic and nosologic criteria for frontotemporal lobar degeneration: consensus of the Consortium for Frontotemporal Lobar Degeneration. *Acta Neuropathologica*, **2007** , 114, 5-22	14.3	837
31	The molecular genetics and neuropathology of frontotemporal lobar degeneration: recent developments. *Neurogenetics*, **2007**, 8, 237-48	3	62
30	The neuropathology of FTD associated With ALS. *Alzheimer Disease and Associated Disorders*, **2007**, 21, S44-9	2.5	50
29	TDP-43 in the ubiquitin pathology of frontotemporal dementia with VCP gene mutations. *Journal of Neuropathology and Experimental Neurology*, **2007**, 66, 152-7	3.1	256
28	A reassessment of the neuropathology of frontotemporal dementia linked to chromosome 3. *Journal of Neuropathology and Experimental Neurology*, **2007**, 66, 884-91	3.1	106
27	TDP-43 in familial and sporadic frontotemporal lobar degeneration with ubiquitin inclusions. *American Journal of Pathology*, **2007**, 171, 227-40	5.8	376
26	TDP-43 in the ubiquitin pathology of frontotemporal dementia with VCP gene mutations. *FASEB Journal*, **2007**, 21, A25	0.9	
25	Familial frontotemporal dementia with neuronal intranuclear inclusions is not a polyglutamine expansion disease. *BMC Neurology*, **2006**, 6, 32	3.1	6
24	Novel ubiquitin neuropathology in frontotemporal dementia with valosin-containing protein gene mutations. *Journal of Neuropathology and Experimental Neurology*, **2006**, 65, 571-81	3.1	182
23	A family with tau-negative frontotemporal dementia and neuronal intranuclear inclusions linked to chromosome 17. *Brain*, **2006**, 129, 853-67	11.2	96
22	The neuropathology of frontotemporal lobar degeneration caused by mutations in the progranulin gene. *Brain*, **2006**, 129, 3081-90	11.2	259
21	Mutations in progranulin explain atypical phenotypes with variants in MAPT. *Brain*, **2006**, 129, 3124-6	11.2	85
20	Mutations in progranulin are a major cause of ubiquitin-positive frontotemporal lobar degeneration. *Human Molecular Genetics*, **2006**, 15, 2988-3001	5.6	463
19	Ubiquitinated TDP-43 in frontotemporal lobar degeneration and amyotrophic lateral sclerosis. *Science*, **2006**, 314, 130-3	33.3	4289

18	Mutations in progranulin cause tau-negative frontotemporal dementia linked to chromosome 17. *Nature*, **2006**, 442, 916-9	50.4	1549
17	Dementia lacking distinctive histology (DLDH) revisited. *Acta Neuropathologica*, **2006**, 112, 551-9	14.3	75
16	Heterogeneity of ubiquitin pathology in frontotemporal lobar degeneration: classification and relation to clinical phenotype. *Acta Neuropathologica*, **2006**, 112, 539-49	14.3	264
15	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. *Journal of Neuropathology and Experimental Neurology*, **2005**, 64, 730-9	3.1	114
14	Neuronal intranuclear inclusions distinguish familial FTD-MND type from sporadic cases. *Dementia and Geriatric Cognitive Disorders*, **2004**, 17, 333-6	2.6	19
13	Extrapyramidal features in patients with motor neuron disease and dementia; a clinicopathological correlative study. *Acta Neuropathologica*, **2004**, 107, 336-40	14.3	19
12	alpha-Internexin aggregates are abundant in neuronal intermediate filament inclusion disease (NIFID) but rare in other neurodegenerative diseases. *Acta Neuropathologica*, **2004**, 108, 213-23	14.3	41
11	alpha-internexin is present in the pathological inclusions of neuronal intermediate filament inclusion disease. *American Journal of Pathology*, **2004**, 164, 2153-61	5.8	106
10	The relationship between extramotor ubiquitin-immunoreactive neuronal inclusions and dementia in motor neuron disease. *Acta Neuropathologica*, **2003**, 105, 98-102	14.3	43
9	Neuronal intranuclear inclusions distinguish familial FTD-MND type from sporadic cases. *Acta Neuropathologica*, **2003**, 105, 543-8	14.3	33
8	Central neurocytoma: histologic atypia, proliferation potential, and clinical outcome. *Cancer*, **1999**, 85, 1606-10	6.4	104
7	Phenotypic variability of Gerstmann-Str□ssler-Scheinker disease is associated with prion protein heterogeneity. *Journal of Neuropathology and Experimental Neurology*, **1998**, 57, 979-88	3.1	157
6	Senile plaques do not progressively accumulate with normal aging. *Acta Neuropathologica*, **1994**, 87, 520-5	14.3	56
5	A lady with weakness, fasciculations, and failing memory90-97		
4	Young man with progressive speech impairment and weakness105-114		
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