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Mutations in progranulin cause tau-negative frontotemporal dementia linked to chromosome 17

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1703	Alzheimer: 100 Years and Beyond. <b>2006</b> ,		5
1702	Ubiquitinated TDP-43 in frontotemporal lobar degeneration and amyotrophic lateral sclerosis. <b>2006</b> , 314, 130-3		4289
1701	Pathological heterogeneity of frontotemporal lobar degeneration with ubiquitin-positive inclusions delineated by ubiquitin immunohistochemistry and novel monoclonal antibodies. <b>2006</b> , 169, 1343-52		266
1700	[Nosology of dementias: the neuropathologist's point of view]. <b>2006</b> , 162, 921-8		8
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1282	Common variant in GRN is a genetic risk factor for hippocampal sclerosis in the elderly. <b>2010</b> , 7, 170-4	71
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1280	Frontotemporal Dementia. <b>2010</b> , 34, 397-416	
1279	Corticobasal Ganglionic Degeneration. <b>2010</b> , 375-396	1
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1270	miR-107 regulates granulin/progranulin with implications for traumatic brain injury and neurodegenerative disease. <b>2010</b> , 177, 334-45	146
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1235	Intra-familial clinical heterogeneity due to FTLD-U with TDP-43 proteinopathy caused by a novel deletion in progranulin gene (PGRN). <b>2010</b> , 22, 1123-33	16
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1183	Mucosal Progranulin expression is induced by <i>H. pylori</i> , but independent of Secretory Leukocyte Protease Inhibitor (SLPI) expression. <b>2011</b> , 11, 63	7

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452	Preferential Disruption of Auditory Word Representations in Primary Progressive Aphasia With the Neuropathology of FTLT-DTP Type A. <b>2019</b> , 32, 46-53	11
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450	Genetic mimics of the non-genetic atypical parkinsonian disorders - the 'atypical' atypical. <b>2019</b> , 149, 327-351	5
449	Parkinsonism in frontotemporal dementias. <b>2019</b> , 149, 249-275	15
448	Characterization of lysosomal proteins Progranulin and Prosaposin and their interactions in Alzheimer's disease and aged brains: increased levels correlate with neuropathology. <b>2019</b> , 7, 215	15
447	Impaired βglucocerebrosidase activity and processing in frontotemporal dementia due to progranulin mutations. <b>2019</b> , 7, 218	24
446	The Western Diet Regulates Hippocampal Microvascular Gene Expression: An Integrated Genomic Analyses in Female Mice. <b>2019</b> , 9, 19058	11
445	Synaptic Pruning by Microglia in Epilepsy. <b>2019</b> , 8,	24

444	Cross-Species Single-Cell Analysis Reveals Divergence of the Primate Microglia Program. <b>2019</b> , 179, 1609-1622.e15	15
443	Review: Modelling the pathology and behaviour of frontotemporal dementia. <b>2019</b> , 45, 58-80	9
442	AAV-Mediated Progranulin Delivery to a Mouse Model of Progranulin Deficiency Causes T Cell-Mediated Toxicity. <b>2019</b> , 27, 465-478	29
441	Peripheral GRN mRNA and Serum Progranulin Levels as a Potential Indicator for Both the Presence of Splice Site Mutations and Individuals at Risk for Frontotemporal Dementia. <b>2019</b> , 67, 159-167	7
440	Dedifferentiated Schwann cells secrete progranulin that enhances the survival and axon growth of motor neurons. <b>2019</b> , 67, 360-375	12
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435	Review: Neuropathology of non-tau frontotemporal lobar degeneration. <b>2019</b> , 45, 19-40	57
434	Rare variants in the neuronal ceroid lipofuscinosis gene MFSD8 are candidate risk factors for frontotemporal dementia. <b>2019</b> , 137, 71-88	20
433	Pathological Changes of Tau Related to Alzheimer's Disease. <b>2019</b> , 10, 931-944	29
432	Neuroinflammation and blood-brain barrier disruption following traumatic brain injury: Pathophysiology and potential therapeutic targets. <b>2020</b> , 98, 19-28	55
431	Altered plasma visfatin levels and insulin resistance in patients with Alzheimer's disease. <b>2020</b> , 120, 901-906	4
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428	The missense p.Trp7Arg mutation in GRN gene leads to progranulin haploinsufficiency. <b>2020</b> , 85, 154.e9-154.e11	11
427	Progranulin deficiency confers resistance to autoimmune encephalomyelitis in mice. <b>2020</b> , 17, 1077-1091	7



426	Anti-GluA3 antibodies in frontotemporal dementia: effects on glutamatergic neurotransmission and synaptic failure. <b>2020</b> , 86, 143-155	16
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423	Approaches to develop therapeutics to treat frontotemporal dementia. <b>2020</b> , 166, 107948	6
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418	One novel GRN null mutation, two different aphasia phenotypes. <b>2020</b> , 87, 141.e9-141.e14	4
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416	International view on genetic frontotemporal dementia. <b>2020</b> , 19, 106-108	
415	Interaction between PGRN gene and the early trauma on clinical characteristics in patients with obsessive-compulsive disorder. <b>2020</b> , 263, 134-140	7
414	A transcriptomic analysis of Nsmce1 overexpression in mouse hippocampal neuronal cell by RNA sequencing. <b>2020</b> , 20, 459-470	2
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412	Invited Review: The role of prion-like mechanisms in neurodegenerative diseases. <b>2020</b> , 46, 522-545	46
411	YB-1 Interferes with TNF $\alpha$ /TNFR Binding and Modulates Progranulin-Mediated Inhibition of TNF $\alpha$ Signaling. <b>2020</b> , 21,	2
410	Neuroimaging in genetic frontotemporal dementia and amyotrophic lateral sclerosis. <b>2020</b> , 145, 105063	10
409	ALS Genetics: Gains, Losses, and Implications for Future Therapies. <b>2020</b> , 108, 822-842	72

408	The complex relationship between genotype, pathology and phenotype in familial dementia. <b>2020</b> , 145, 105082	1
407	Secreted Chaperones in Neurodegeneration. <b>2020</b> , 12, 268	11
406	The Rise of the GRN C157KfsX97 Mutation in Southern Italy: Going Back to the Fall of the Western Roman Empire. <b>2020</b> , 78, 387-394	1
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404	Phase Separation and Neurodegenerative Diseases: A Disturbance in the Force. <b>2020</b> , 55, 45-68	70
403	Molecular and cellular mechanisms underlying the pathogenesis of Alzheimer's disease. <b>2020</b> , 15, 40	165
402	Serum progranulin levels in paediatric patients with Gaucher disease; relation to disease severity and liver stiffness by transient elastography. <b>2020</b> , 40, 3051-3060	1
401	Elevated levels of extracellular vesicles in progranulin-deficient mice and FTD-GRN Patients. <b>2020</b> , 7, 2433-2449	4
400	Longitudinal Characterization of Transcriptomic, Functional, and Morphological Features in Human iPSC-Derived Neurons and Their Application to Investigate Translational Progranulin Disease Biology. <b>2020</b> , 12, 576678	1
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379	Reappraisal of the anatomical spreading and propagation hypothesis about TDP-43 aggregation in amyotrophic lateral sclerosis and frontotemporal lobar degeneration. <b>2020</b> , 40, 426-435	8
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377	Impairment of Lysosome Function and Autophagy in Rare Neurodegenerative Diseases. <b>2020</b> , 432, 2714-2734	23
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373	Altered levels of CSF proteins in patients with FTD, presymptomatic mutation carriers and non-carriers. <b>2020</b> , 9, 27	10

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367	Genetic Diversity in Frontotemporal Dementia. <b>2020</b> , 54, 13-23	5
366	Pathomechanism Heterogeneity in the Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Disease Spectrum: Providing Focus Through the Lens of Autophagy. <b>2020</b> , 432, 2692-2713	11
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362	Comparison of sporadic and familial behavioral variant frontotemporal dementia (FTD) in a North American cohort. <b>2020</b> , 16, 60-70	17
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358	Experimental gene therapies for the NCLs. <b>2020</b> , 1866, 165772	5
357	Frontotemporal Dementia: Neuropathology, Genetics, Neuroimaging, and Treatments. <b>2020</b> , 43, 331-344	10
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355	Convulsive responses to seizure-inducible drugs are exacerbated in progranulin-deficient mice. <b>2020</b> , 31, 478-483	1

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342	Clinical and Neuroimaging Aspects of Familial Frontotemporal Lobar Degeneration Associated with MAPT and GRN Mutations. <b>2021</b> , 1281, 77-92	0
341	Reply: Two heterozygous progranulin mutations in progressive supranuclear palsy. <b>2021</b> , 144, e28	
340	Differential regulation of progranulin derived granulin peptides.	1
339	The Right Temporal Variant of Frontotemporal Dementia Is Not Genetically Sporadic: A Case Series. <b>2021</b> , 79, 1195-1201	5
338	CRISPR/Cas9-mediated grna gene knockout leads to neurodevelopmental defects and motor behavior changes in zebrafish. <b>2021</b> , 157, 520-531	3
337	Outcomes of Progranulin Gene Therapy in the Retina are Dependent on Time of Delivery.	

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334	Defective Lysosomal Lipid Catabolism as a Common Pathogenic Mechanism for Dementia. <b>2021</b> , 23, 1-24	3
333	Genetics of frontotemporal dementia in China. <b>2021</b> , 22, 321-335	4
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322	Neuropathological and behavioral characterization of aged Grn R493X progranulin-deficient frontotemporal dementia knockin mice. <b>2021</b> , 9, 57	2
321	Structural insight into mutations at 155 position of valosin containing protein (VCP) linked to inclusion body myopathy with Paget disease of bone and frontotemporal Dementia. <b>2021</b> , 28, 2128-2138	
320	Autoantibodies against Progranulin and IL-1 receptor antagonist in critically ill COVID-19.	1
319	Quantitative endophenotypes as an alternative approach to understanding genetic risk in neurodegenerative diseases. <b>2021</b> , 151, 105247	1

318	Whole-Exome Sequencing and C9orf72 Analysis in Primary Progressive Aphasia. <b>2021</b> , 80, 985-990	3
317	Association of Hemoglobin A1C With TDP-43 Pathology in Community-Based Elders. <b>2021</b> ,	0
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315	Gene Expression Imputation Across Multiple Tissue Types Provides Insight Into the Genetic Architecture of Frontotemporal Dementia and Its Clinical Subtypes. <b>2021</b> , 89, 825-835	3
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303	Progranulin deficiency in Iba-1 myeloid cells exacerbates choroidal neovascularization by perturbation of lysosomal function and abnormal inflammation. <b>2021</b> , 18, 164	4
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300	Tweaking Progranulin Expression: Therapeutic Avenues and Opportunities. <b>2021</b> , 14, 713031	3
299	Neuronal VCP loss of function recapitulates FTLD-TDP pathology. <b>2021</b> , 36, 109399	5
298	Loss of TREM2 reduces hyperactivation of progranulin deficient microglia but not lysosomal pathology.	1
297	FTLD Patient-Derived Fibroblasts Show Defective Mitochondrial Function and Accumulation of p62. <b>2021</b> , 58, 5438-5458	1
296	TDP-43 and Inflammation: Implications for Amyotrophic Lateral Sclerosis and Frontotemporal Dementia. <b>2021</b> , 22,	4
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292	Progranulin associates with Rab2 and is involved in autophagosome-lysosome fusion in Gaucher disease. <b>2021</b> , 99, 1639-1654	1
291	Processing of progranulin into granulins involves multiple lysosomal proteases and is affected in frontotemporal lobar degeneration. <b>2021</b> , 16, 51	4
290	Deciphering Neurodegenerative Diseases Using Long-Read Sequencing. <b>2021</b> , 97, 423-433	1
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288	Plasma NfL levels and longitudinal change rates in and -associated diseases: from tailored references to clinical applications. <b>2021</b> , 92, 1278-1288	2
287	Prediction and verification of the AD-FTLD common pathomechanism based on dynamic molecular network analysis. <b>2021</b> , 4, 961	1
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285	Mice Treated Subcutaneously with Mouse LPS-Converted PrP or LPS Alone Showed Brain Gene Expression Profiles Characteristic of Prion Disease. <b>2021</b> , 8,	
284	Molecular Pathways Involved in Frontotemporal Lobar Degeneration with TDP-43 Proteinopathy: What Can We Learn from Proteomics?. <b>2021</b> , 22,	1
283	Rescue of a lysosomal storage disorder caused by Grn loss of function with a brain penetrant progranulin biologic. <b>2021</b> , 184, 4651-4668.e25	11



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281	Outcomes of progranulin gene therapy in the retina are dependent on time and route of delivery. <b>2021</b> , 22, 40-51	1
280	Effect of the Histone Deacetylase Inhibitor FRM-0334 on Progranulin Levels in Patients With Progranulin Gene Haploinsufficiency: A Randomized Clinical Trial. <b>2021</b> , 4, e2125584	2
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278	Progranulin deficiency results in reduced bis(monoacylglycero)phosphate (BMP) levels and gangliosidosis.	0
277	Genetic variants in progranulin upstream open reading frames increase downstream protein expression. <b>2021</b> , 110, 113-113	
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274	Abnormal spatiotemporal expression pattern of progranulin and neurodevelopment impairment in VPA-induced ASD rat model. <b>2021</b> , 196, 108689	3
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272	Genome-wide association study of frontotemporal dementia identifies a C9ORF72 haplotype with a median of 12-G4C2 repeats that predisposes to pathological repeat expansions. <b>2021</b> , 11, 451	0
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270	Glucocerebrosidase mutations: A paradigm for neurodegeneration pathways. <b>2021</b> , 175, 42-55	1
269	Younger-Onset Dementias: Behavioral Neurology/Brain Diseases/Healthy & Pathological Aging. <b>2022</b> , 88-96	
268	Fluid Biomarkers of Frontotemporal Lobar Degeneration. <b>2021</b> , 1281, 123-139	1
267	FDG-PET in presymptomatic C9orf72 mutation carriers. <b>2021</b> , 31, 102687	2
266	Disease-related cortical thinning in presymptomatic granulin mutation carriers. <b>2021</b> , 29, 102540	2
265	Fibroblasts from idiopathic Parkinson's disease exhibit deficiency of lysosomal glucocerebrosidase activity associated with reduced levels of the trafficking receptor LIMP2. <b>2021</b> , 14, 16	5

264	Knowledge assessment and psychological impact of genetic counseling in people at risk for familial FTD. <b>2021</b> , 13, e12225	1
263	Single-cell deconvolution of 3,000 post-mortem brain samples for eQTL and GWAS dissection in mental disorders.	4
262	TDP-43 y su incidencia en demencias degenerativas. <b>2021</b> , 13, 37-47	
261	SLITRK2, an X-linked modifier of the age at onset in C9orf72 frontotemporal lobar degeneration. <b>2021</b> , 144, 2798-2811	2
260	Molecular Genetics of Frontotemporal Dementia.	2
259	Frontotemporal Dementia and Parkinsonism Linked to Chromosome 17. 110-134	20
258	Frontotemporal Lobar Degeneration with TDP-43 Pathology. 393-403	1
257	Neuropsychology of Movement Disorders and Motor Neuron Disease. <b>2010</b> , 315-333	2
256	Animal Models of Amyotrophic Lateral Sclerosis. <b>2011</b> , 515-531	1
255	The Ubiquitin System in Alzheimer's Disease. <b>2020</b> , 1233, 195-221	25
254	Genetik und Gen-Umwelt-Interaktionen bei psychischen Erkrankungen. <b>2017</b> , 147-191	2
253	Altered lipid metabolic homeostasis in the pathogenesis of Alzheimer's disease. <b>2020</b> , 469-504	3
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251	Disorders of Upper and Lower Motor Neurons. <b>2008</b> , 2183-2220	3
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