

JÃ,rgen Erik Nielsen

List of Publications by Year in descending order

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Version: 2024-02-01

65
papers

2,337
citations

393982

19
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223531

46
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72
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72
docs citations

72
times ranked

4415
citing authors

#	ARTICLE	IF	CITATIONS
1	Six generations of <i>CHMP2B</i> -mediated Frontotemporal Dementia: Clinical features, predictive testing, progression, and survival. <i>Acta Neurologica Scandinavica</i> , 2022, 145, 529-540.	1.0	4
2	Peripheral helper T cells in the pathogenesis of multiple sclerosis. <i>Multiple Sclerosis Journal</i> , 2022, 28, 1340-1350.	1.4	3
3	Widening the spectrum of spinocerebellar ataxia autosomal recessive type 10 (SCAR10). <i>BMJ Case Reports</i> , 2022, 15, e248228.	0.2	1
4	Impairments of social cognition significantly predict the progression of functional decline in Huntington's disease: A 6-year follow-up study. <i>Applied Neuropsychology Adult</i> , 2022, , 1-10.	0.7	3
5	Decreased CSF oxytocin relates to measures of social cognitive impairment in Huntington's disease patients. <i>Parkinsonism and Related Disorders</i> , 2022, 99, 23-29.	1.1	8
6	Increased Intrathecal Activity of Follicular Helper T Cells in Patients With Relapsing-Remitting Multiple Sclerosis. <i>Neurology: Neuroimmunology and Neuroinflammation</i> , 2022, 9, .	3.1	11
7	Novel Homozygous Truncating Variant Widens the Spectrum of Early-Onset Multisystemic SYNE1 Ataxia. <i>Cerebellum</i> , 2021, , 1.	1.4	0
8	Endophenotypical drift in Huntington's disease: a 5-year follow-up study. <i>Orphanet Journal of Rare Diseases</i> , 2021, 16, 340.	1.2	3
9	F21...On the association between apathy and deficits of social cognition and executive functions in huntington's disease. , 2021, , .		0
10	CCG-CCG interruptions in high-penetrance SCA8 families increase RAN translation and protein toxicity. <i>EMBO Molecular Medicine</i> , 2021, 13, e14095.	3.3	12
11	Astrocytic reactivity triggered by defective autophagy and metabolic failure causes neurotoxicity in frontotemporal dementia type 3. <i>Stem Cell Reports</i> , 2021, 16, 2736-2751.	2.3	23
12	Cortical Frontoparietal Network Dysfunction in -Frontotemporal Dementia. <i>Frontiers in Aging Neuroscience</i> , 2021, 13, 714220.	1.7	0
13	Intellectual Curiosity and Action Initiation are Subtypes of Apathy Affected in Huntington Disease Gene Expansion Carriers. <i>Cognitive and Behavioral Neurology</i> , 2021, 34, 295-302.	0.5	2
14	Intellectual curiosity and action initiation are subtypes of apathy affected in Huntington's disease gene expansion carriers. <i>Alzheimer's and Dementia</i> , 2021, 17, .	0.4	0
15	Early Intrathecal T Helper 17.1 Cell Activity in Huntington Disease. <i>Annals of Neurology</i> , 2020, 87, 246-255.	2.8	24
16	Enhancement of Autophagy and Solubilization of Ataxin-2 Alleviate Apoptosis in Spinocerebellar Ataxia Type 2 Patient Cells. <i>Cerebellum</i> , 2020, 19, 165-181.	1.4	11
17	Mania triggered by levodopa treatment in a patient with frontotemporal dementia caused by a C9orf72 repeat expansion: A case report. <i>Clinical Neurology and Neurosurgery</i> , 2020, 198, 106147.	0.6	3
18	Paroxysmal Cranial Dyskinesia and Nail-Patella Syndrome Caused by a Novel Variant in the LMX1B Gene. <i>Movement Disorders</i> , 2020, 35, 2343-2347.	2.2	2

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19	Glutamate-glutamine homeostasis is perturbed in neurons and astrocytes derived from patient iPSC models of frontotemporal dementia. <i>Molecular Brain</i> , 2020, 13, 125.	1.3	36
20	Peripheral neuropathy in hereditary spastic paraplegia caused by REEP1 variants. <i>Journal of Neurology</i> , 2019, 266, 735-744.	1.8	11
21	Genotype-phenotype correlations, dystonia and disease progression in spinocerebellar ataxia type 14. <i>Movement Disorders</i> , 2018, 33, 1119-1129.	2.2	26
22	Beneficial effect of intravenous immunoglobulin treatment in a patient with antiphospholipid syndrome associated chorea. <i>Journal of the Neurological Sciences</i> , 2018, 390, 52-53.	0.3	2
23	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.	4.9	97
24	CSF neurofilament light concentration is increased in presymptomatic <i>CHMP2B</i> mutation carriers. <i>Neurology</i> , 2018, 90, e157-e163.	1.5	11
25	Quantitative Measurements of Motor Function in Alzheimer's Disease, Frontotemporal Dementia, and Dementia with Lewy Bodies: A Proof-of-Concept Study. <i>Dementia and Geriatric Cognitive Disorders</i> , 2018, 46, 168-179.	0.7	9
26	Inflammatory markers of CHMP2B-mediated frontotemporal dementia. <i>Journal of Neuroimmunology</i> , 2018, 324, 136-142.	1.1	10
27	Sporadic Creutzfeldt-Jakob Disease in a Woman Married Into a Gerstmann-StrÄussler-Scheinker Family: An Investigation of Prions Transmission via Microchimerism. <i>Journal of Neuropathology and Experimental Neurology</i> , 2018, 77, 673-684.	0.9	6
28	D03...Quality control for plasma and cerebrospinal fluid samples using mass spectrometry. , 2018, , .		0
29	SCA28: Novel Mutation in the AFG3L2 Proteolytic Domain Causes a Mild Cerebellar Syndrome with Selective Type-1 Muscle Fiber Atrophy. <i>Cerebellum</i> , 2017, 16, 62-67.	1.4	16
30	A Novel TTBK2 De Novo Mutation in a Danish Family with Early-Onset Spinocerebellar Ataxia. <i>Cerebellum</i> , 2017, 16, 268-271.	1.4	15
31	Patient iPSC-Derived Neurons for Disease Modeling of Frontotemporal Dementia with Mutation in CHMP2B. <i>Stem Cell Reports</i> , 2017, 8, 648-658.	2.3	65
32	Characterization of energy and neurotransmitter metabolism in cortical glutamatergic neurons derived from human induced pluripotent stem cells: A novel approach to study metabolism in human neurons. <i>Neurochemistry International</i> , 2017, 106, 48-61.	1.9	14
33	Hereditary cerebral small vessel disease and stroke. <i>Clinical Neurology and Neurosurgery</i> , 2017, 155, 45-57.	0.6	62
34	Evidence of oxidative stress and mitochondrial dysfunction in spinocerebellar ataxia type 2 (SCA2) patient fibroblasts: Effect of coenzyme Q10 supplementation on these parameters. <i>Mitochondrion</i> , 2017, 34, 103-114.	1.6	42
35	Defining active progressive multiple sclerosis. <i>Multiple Sclerosis Journal</i> , 2017, 23, 1727-1735.	1.4	34
36	TMEM106B and ApoE polymorphisms in CHMP2B-mediated frontotemporal dementia (FTD-3). <i>Neurobiology of Aging</i> , 2017, 59, 221.e1-221.e7.	1.5	4

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37	Neurons derived from sporadic Alzheimer's disease iPSCs reveal elevated TAU hyperphosphorylation, increased amyloid levels, and GSK3B activation. <i>Alzheimer's Research and Therapy</i> , 2017, 9, 90.	3.0	161
38	Generation of spinocerebellar ataxia type 2 patient-derived iPSC line H196. <i>Stem Cell Research</i> , 2016, 16, 199-201.	0.3	4
39	Liver function in Huntington's disease assessed by blood biochemical analyses in a clinical setting. <i>Journal of the Neurological Sciences</i> , 2016, 362, 326-332.	0.3	12
40	Generation of an isogenic, gene-corrected control cell line of the spinocerebellar ataxia type 2 patient-derived iPSC line H196. <i>Stem Cell Research</i> , 2016, 16, 162-165.	0.3	10
41	Induced pluripotent stem cell - derived neurons for the study of spinocerebellar ataxia type 3. <i>Stem Cell Research</i> , 2016, 17, 306-317.	0.3	27
42	Selected CSF biomarkers indicate no evidence of early neuroinflammation in Huntington disease. <i>Neurology: Neuroimmunology and NeuroInflammation</i> , 2016, 3, e287.	3.1	53
43	Personality traits in Huntington's disease: An exploratory study of gene expansion carriers and non-carriers. <i>American Journal of Medical Genetics Part B: Neuropsychiatric Genetics</i> , 2016, 171, 1153-1160.	1.1	6
44	Generation of an isogenic, gene-corrected control cell line of the spinocerebellar ataxia type 2 patient-derived iPSC line H271. <i>Stem Cell Research</i> , 2016, 16, 180-183.	0.3	15
45	Generation of a human induced pluripotent stem cell line via CRISPR-Cas9 mediated integration of a site-specific homozygous mutation in CHMP2B. <i>Stem Cell Research</i> , 2016, 17, 151-153.	0.3	5
46	Generation of a human induced pluripotent stem cell line via CRISPR-Cas9 mediated integration of a site-specific heterozygous mutation in CHMP2B. <i>Stem Cell Research</i> , 2016, 17, 148-150.	0.3	6
47	Do I misconstrue? Sarcasm detection, emotion recognition, and theory of mind in Huntington disease.. <i>Neuropsychology</i> , 2016, 30, 181-189.	1.0	39
48	Generation of spinocerebellar ataxia type 2 patient-derived iPSC line H271. <i>Stem Cell Research</i> , 2016, 16, 159-161.	0.3	3
49	Generation of an isogenic, gene-corrected control cell line of the spinocerebellar ataxia type 2 patient-derived iPSC line H266. <i>Stem Cell Research</i> , 2016, 16, 202-205.	0.3	17
50	Generation of spinocerebellar ataxia type 2 patient-derived iPSC line H266. <i>Stem Cell Research</i> , 2016, 16, 166-169.	0.3	3
51	Generation of spinocerebellar ataxia type 3 patient-derived induced pluripotent stem cell line SCA3.A11. <i>Stem Cell Research</i> , 2016, 16, 553-556.	0.3	7
52	Generation of spinocerebellar ataxia type 3 patient-derived induced pluripotent stem cell line SCA3.B11. <i>Stem Cell Research</i> , 2016, 16, 589-592.	0.3	9
53	Social Cognition, Executive Functions and Self-Report of Psychological Distress in Huntington's Disease. <i>PLOS Currents</i> , 2016, 8, .	1.4	7
54	Frontotemporal dementia caused by CHMP2B mutation is characterised by neuronal lysosomal storage pathology. <i>Acta Neuropathologica</i> , 2015, 130, 511-523.	3.9	79

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55	Assessing Impairment of Executive Function and Psychomotor Speed in Premanifest and Manifest Huntington's Disease Gene-expansion Carriers. <i>Journal of the International Neuropsychological Society</i> , 2015, 21, 193-202.	1.2	25
56	Exploring Genetic Factors Involved in Huntington Disease Age of Onset: E2F2 as a New Potential Modifier Gene. <i>PLoS ONE</i> , 2015, 10, e0131573.	1.1	11
57	Reduced ceramide synthase 2 activity causes progressive myoclonic epilepsy. <i>Annals of Clinical and Translational Neurology</i> , 2014, 1, 88-98.	1.7	50
58	Epigenetic remodelling and dysregulation of DLGAP4 is linked with early-onset cerebellar ataxia. <i>Human Molecular Genetics</i> , 2014, 23, 6163-6176.	1.4	19
59	Reduction in mitochondrial DNA copy number in peripheral leukocytes after onset of Huntington's disease. <i>Mitochondrion</i> , 2014, 17, 14-21.	1.6	54
60	YKL-40 in cerebrospinal fluid in Huntington's disease – A role in pathology or a nonspecific response to inflammation?. <i>Parkinsonism and Related Disorders</i> , 2014, 20, 1301-1303.	1.1	20
61	A clinical classification acknowledging neuropsychiatric and cognitive impairment in Huntington's disease. <i>Orphanet Journal of Rare Diseases</i> , 2014, 9, 114.	1.2	50
62	Frontotemporal dementia and its subtypes: a genome-wide association study. <i>Lancet Neurology</i> , The, 2014, 13, 686-699.	4.9	302
63	Antisense Gene Silencing: Therapy for Neurodegenerative Disorders?. <i>Genes</i> , 2013, 4, 457-484.	1.0	14
64	ATXN2 with intermediate-length CAG/CAA repeats does not seem to be a risk factor in hereditary spastic paraplegia. <i>Journal of the Neurological Sciences</i> , 2012, 321, 100-102.	0.3	7
65	Mutations in the endosomal ESCRTIII-complex subunit CHMP2B in frontotemporal dementia. <i>Nature Genetics</i> , 2005, 37, 806-808.	9.4	752