Conrad C Weihl

List of Publications by Year
in descending order

70 papers	9,258 citations	87888 38 h-index	82547 72 g-index
78 all docs	78 docs citations	78 times ranked	16641 citing authors

#	Article	IF	Citations
1	Current and Future Approaches to Classify VUSs in LGMD-Related Genes. Genes, 2022, 13, 382.	2.4	3
2	Randomized phase 2 study of <scp>ACE</scp>â€083, a <scp>muscleâ€promoting</scp> agent, in facioscapulohumeral muscular dystrophy. Muscle and Nerve, 2022, 66, 50-62.	2.2	8
3	VCP suppresses proteopathic seeding in neurons. Molecular Neurodegeneration, 2022, 17, 30.	10.8	15
4	Progression to Loss of Ambulation Among Patients with Autosomal Recessive Limb-girdle Muscular Dystrophy: A Systematic Review. Journal of Neuromuscular Diseases, 2022, , 1-16.	2.6	3
5	Multisystem Proteinopathy Due to VCP Mutations: A Review of Clinical Heterogeneity and Genetic Diagnosis. Genes, 2022, 13, 963.	2.4	28
6	Clinical utility of antiâ€cytosolic 5â€™â€nucleotidase 1A antibody in idiopathic inflammatory myopathies. Annals of Clinical and Translational Neurology, 2021, 8, 571-578.	3.7	18
7	Neuronal VCP loss of function recapitulates FTLD-TDP pathology. Cell Reports, 2021, 36, 109399.	6.4	25
8	Molecular and cellular basis of genetically inherited skeletal muscle disorders. Nature Reviews Molecular Cell Biology, 2021, 22, 713-732.	37.0	55
9	Loss-of-function mutation in VCP mimics the characteristic pathology as in FTLD-TARDBP. Autophagy, 2021, 17, 4502-4503.	9.1	5
10	Mutations in the J domain of DNAJB6 cause dominant distal myopathy. Neuromuscular Disorders, 2020, 30, 38-46.	0.6	20
11	A cross-sectional study of hand function in inclusion body myositis: Implications for functional rating scale. Neuromuscular Disorders, 2020, 30, 200-206.	0.6	4
12	Optimizing hand-function patient outcome measures for inclusion body myositis. Neuromuscular Disorders, 2020, 30, 807-814.	0.6	5
13	Client processing is altered by novel myopathy-causing mutations in the HSP40 J domain. PLoS ONE, 2020, 15, e0234207.	2.5	6
14	High-protein diets increase cardiovascular risk by activating macrophage mTOR to suppress mitophagy. Nature Metabolism, 2020, 2, 110-125.	11.9	85
15	Inhibition of DNAJ-HSP70 interaction improves strength in muscular dystrophy. Journal of Clinical Investigation, 2020, 130, 4470-4485.	8.2	14
16	Phenotypic diversity in an international Cure VCP Disease registry. Orphanet Journal of Rare Diseases, 2020, 15, 267.	2.7	11
17	Loss of lipin 1â€mediated phosphatidic acid phosphohydrolase activity in muscle leads to skeletal myopathy in mice. FASEB Journal, 2019, 33, 652-667.	0.5	30
18	Desmin forms toxic, seeding-competent amyloid aggregates that persist in muscle fibers. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 16835-16840.	7.1	36

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19	Limbâ€girdle muscular dystrophy: A perspective from adult patients on what matters most. Muscle and Nerve, 2019, 60, 419-424.	2.2	15
20	VCP maintains lysosomal homeostasis and TFEB activity in differentiated skeletal muscle. Autophagy, 2019, 15, 1082-1099.	9.1	44
21	Estimating prevalence for limb-girdle muscular dystrophy based on public sequencing databases. Genetics in Medicine, 2019, 21, 2512-2520.	2.4	56
22	Lithium chloride corrects weakness and myopathology in a preclinical model of LGMD1D. Neurology: Genetics, 2019, 5, e318.	1.9	15
23	A metastable subproteome underlies inclusion formation in muscle proteinopathies. Acta Neuropathologica Communications, 2019, 7, 197.	5.2	16
24	Sporadic Inclusion Body Myositis and Other Rimmed Vacuolar Myopathies. CONTINUUM Lifelong Learning in Neurology, 2019, 25, 1586-1598.	0.8	6
25	CANOMAD and other chronic ataxic neuropathies with disialosyl antibodies (CANDA). Journal of Neurology, 2018, 265, 1402-1409.	3.6	40
26	Myopathy associated BAG3 mutations lead to protein aggregation by stalling Hsp70 networks. Nature Communications, 2018, 9, 5342.	12.8	65
27	Homozygous recessive MYH2 mutation mimicking dominant MYH2 associated myopathy. Neuromuscular Disorders, 2018, 28, 675-679.	0.6	10
28	TIA1 variant drives myodegeneration in multisystem proteinopathy with SQSTM1 mutations. Journal of Clinical Investigation, 2018, 128, 1164-1177.	8.2	75
29	Establishing prevalence in rare neuromuscular diseases. Neurology: Genetics, 2017, 3, e146.	1.9	1
30	Quantification of autophagy flux using LC3 ELISA. Analytical Biochemistry, 2017, 530, 57-67.	2.4	43
31	Cystinosis distal myopathy, novel clinical, pathological and genetic features. Neuromuscular Disorders, 2017, 27, 873-878.	0.6	5
32	Exploiting macrophage autophagy-lysosomal biogenesis as a therapy for atherosclerosis. Nature Communications, 2017, 8, 15750.	12.8	258
33	Keap1/Cullin3 Modulates p62/SQSTM1 Activity via UBA Domain Ubiquitination. Cell Reports, 2017, 19, 188-202.	6.4	110
34	Proteomics of rimmed vacuoles define new risk allele in inclusion body myositis. Annals of Neurology, 2017, 81, 227-239.	5.3	59
35	Comparisons of ELISA and Western blot assays for detection of autophagy flux. Data in Brief, 2017, 13, 696-699.	1.0	8
36	Regulation of SQSTM1/p62 via UBA domain ubiquitination and its role in disease. Autophagy, 2017, 13, 1615-1616.	9.1	47

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37	<scp>VCP</scp> /p97 cooperates with <scp>YOD</scp> 1, <scp>UBXD</scp> 1 and <scp>PLAA</scp> to drive clearance of ruptured lysosomes by autophagy. EMBO Journal, 2017, 36, 135-150.	7.8	259
38	215th ENMC International Workshop VCP-related multi-system proteinopathy (IBMPFD) 13â€“15 November 2015, Heemskerk, The Netherlands. Neuromuscular Disorders, 2016, 26, 535-547.	0.6	38
39	Rare Manifestation of a c.290 C>T, p.Gly97GluVCPMutation. Case Reports in Genetics, 2015, 2015, 1-5.	0.2	16
40	Autophagic vacuolar pathology in desminopathies. Neuromuscular Disorders, 2015, 25, 199-206.	0.6	19
41	Targeted sequencing and identification of genetic variants in sporadic inclusion body myositis. Neuromuscular Disorders, 2015, 25, 289-296.	0.6	56
42	Altered cofactor regulation with disease-associated p97/VCP mutations. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, E1705-14.	7.1	87
43	SQSTM1 splice site mutation in distal myopathy with rimmed vacuoles. Neurology, 2015, 85, 665-674.	1.1	74
44	Myofibrillar disruption and RNA-binding protein aggregation in a mouse model of limb-girdle muscular dystrophy 1D. Human Molecular Genetics, 2015, 24, 6588-6602.	2.9	31
45	Myopathy-causing Mutations in an HSP40 Chaperone Disrupt Processing of Specific Client Conformers. Journal of Biological Chemistry, 2014, 289, 21120-21130.	3.4	46
46	Prion-like nuclear aggregation of TDP-43 during heat shock is regulated by HSP40/70 chaperones. Human Molecular Genetics, 2014, 23, 157-170.	2.9	102
47	Mutations in the Matrin 3 gene cause familial amyotrophic lateral sclerosis. Nature Neuroscience, 2014, 17, 664-666.	14.8	398
48	The VCP/p97 system at a glance: connecting cellular function to disease pathogenesis. Journal of Cell Science, 2014, 127, 3877-83.	2.0	329
49	A novel mutation in VCP causes Charcotâ€“Marieâ€“Tooth Type 2 disease. Brain, 2014, 137, 2897-2902.	7.6	116
50	Specific Inhibition of p97/VCP ATPase and Kinetic Analysis Demonstrate Interaction between D1 and D2 ATPase Domains. Journal of Molecular Biology, 2014, 426, 2886-2899.	4.2	103
51	Monitoring Autophagy in the Treatment of Protein Aggregate Diseases: Steps Toward Identifying Autophagic Biomarkers. Neurotherapeutics, 2013, 10, 383-390.	4.4	11
52	Mutations in prion-like domains in hnRNPA2B1 and hnRNPA1 cause multisystem proteinopathy and ALS. Nature, 2013, 495, 467-473.	27.8	1,249
53	Motor neuron involvement in multisystem proteinopathy. Neurology, 2013, 80, 1874-1880.	1.1	85
54	mTOR dysfunction contributes to vacuolar pathology and weakness in valosin-containing protein associated inclusion body myopathy. Human Molecular Genetics, 2013, 22, 1167-1179.	2.9	58

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55	Increased autophagy accelerates colchicine-induced muscle toxicity. Autophagy, 2013, 9, 2115-2125.	9.1	35
56	Guidelines for the use and interpretation of assays for monitoring autophagy. Autophagy, 2012, 8, 445-544.	9.1	3,122
57	Exome sequencing reveals DNAJB6 mutations in dominantlyâ€inherited myopathy. Annals of Neurology, 2012, 71, 407-416.	5.3	148
58	Mdx mice have a defect in autophagy that is restored by rapamycinâ€loaded nanoparticle treatment. FASEB Journal, 2012, 26, 396.5.	0.5	1
59	Novel GNE mutations in two phenotypically distinct HIBM2 patients. Neuromuscular Disorders, 2011, 21, 102-105.	0.6	15
60	Endolysosomal sorting of ubiquitylated caveolin-1 is regulated by VCP and UBXD1 and impaired by VCP disease mutations. Nature Cell Biology, 2011, 13, 1116-1123.	10.3	213
61	Another VCP interactor: NF is enough. Journal of Clinical Investigation, 2011, 121, 4627-4630.	8.2	7
62	Sporadic inclusion body myositis: possible pathogenesis inferred from biomarkers. Current Opinion in Neurology, 2010, 23, 482-488.	3.6	47
63	Interaction with Polyglutamine Aggregates Reveals a Q/N-rich Domain in TDP-43. Journal of Biological Chemistry, 2010, 285, 26304-26314.	3.4	138
64	Valosin-containing protein (VCP) is required for autophagy and is disrupted in VCP disease. Journal of Cell Biology, 2009, 187, 875-888.	5.2	444
65	Valosin-containing protein disease: Inclusion body myopathy with Pagetâ€™s disease of the bone and fronto-temporal dementia. Neuromuscular Disorders, 2009, 19, 308-315.	0.6	205
66	Inflammatory myopathies with mitochondrial pathology and protein aggregates. Journal of the Neurological Sciences, 2009, 278, 25-29.	0.6	91
67	Transgenic expression of inclusion body myopathy associated mutant p97/VCP causes weakness and ubiquitinated protein inclusions in mice. Human Molecular Genetics, 2007, 16, 919-928.	2.9	99
68	Motor neuron disease associated with copper deficiency. Muscle and Nerve, 2006, 34, 789-793.	2.2	76
69	Valproate may improve strength and function in patients with type III/IV spinal muscle atrophy. Neurology, 2006, 67, 500-501.	1.1	111
70	Inclusion body myopathy-associated mutations in p97/VCP impair endoplasmic reticulum-associated degradation. Human Molecular Genetics, 2006, 15, 189-199.	2.9	169