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	Guidelines for the use and interpretation of assays for monitoring autophagy. Autophagy, 2012, 8, 445-544.	9.1	3,122
2	Mutations in prion-like domains in hnRNPA2B1 and hnRNPA1 cause multisystem proteinopathy and ALS. Nature, 2013, 495, 467-473.	27.8	1,249
3	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
4	Mutations in the Matrin 3 gene cause familial amyotrophic lateral sclerosis. Nature Neuroscience, 2014, 17, 664-666.	14.8	398
5	The VCP/p97 system at a glance: connecting cellular function to disease pathogenesis. Journal of Cell Science, 2014, 127, 3877-83.	2.0	329
6	<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
7	Exploiting macrophage autophagy-lysosomal biogenesis as a therapy for atherosclerosis. Nature Communications, 2017, 8, 15750.	12.8	258
8	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
9	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
10	Inclusion body myopathy-associated mutations in p97/VCP impair endoplasmic reticulum-associated degradation. Human Molecular Genetics, 2006, 15, 189-199.	2.9	169
11	Exome sequencing reveals DNAJB6 mutations in dominantlyâ€inherited myopathy. Annals of Neurology, 2012, 71, 407-416.	5.3	148
12	Interaction with Polyglutamine Aggregates Reveals a Q/N-rich Domain in TDP-43. Journal of Biological Chemistry, 2010, 285, 26304-26314.	3.4	138
13	A novel mutation in VCP causes Charcotâ€“Marieâ€“Tooth Type 2 disease. Brain, 2014, 137, 2897-2902.	7.6	116
14	Valproate may improve strength and function in patients with type III/IV spinal muscle atrophy. Neurology, 2006, 67, 500-501.	1.1	111
15	Keap1/Cullin3 Modulates p62/SQSTM1 Activity via UBA Domain Ubiquitination. Cell Reports, 2017, 19, 188-202.	6.4	110
16	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
17	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
18	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

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19	Inflammatory myopathies with mitochondrial pathology and protein aggregates. Journal of the Neurological Sciences, 2009, 278, 25-29.	0.6	91
20	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
21	Motor neuron involvement in multisystem proteinopathy. Neurology, 2013, 80, 1874-1880.	1.1	85
22	High-protein diets increase cardiovascular risk by activating macrophage mTOR to suppress mitophagy. Nature Metabolism, 2020, 2, 110-125.	11.9	85
23	Motor neuron disease associated with copper deficiency. Muscle and Nerve, 2006, 34, 789-793.	2.2	76
24	TIA1 variant drives myodegeneration in multisystem proteinopathy with SQSTM1 mutations. Journal of Clinical Investigation, 2018, 128, 1164-1177.	8.2	75
25	SQSTM1 splice site mutation in distal myopathy with rimmed vacuoles. Neurology, 2015, 85, 665-674.	1.1	74
26	Myopathy associated BAG3 mutations lead to protein aggregation by stalling Hsp70 networks. Nature Communications, 2018, 9, 5342.	12.8	65
27	Proteomics of rimmed vacuoles define new risk allele in inclusion body myositis. Annals of Neurology, 2017, 81, 227-239.	5.3	59
28	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
29	Targeted sequencing and identification of genetic variants in sporadic inclusion body myositis. Neuromuscular Disorders, 2015, 25, 289-296.	0.6	56
30	Estimating prevalence for limb-girdle muscular dystrophy based on public sequencing databases. Genetics in Medicine, 2019, 21, 2512-2520.	2.4	56
31	Molecular and cellular basis of genetically inherited skeletal muscle disorders. Nature Reviews Molecular Cell Biology, 2021, 22, 713-732.	37.0	55
32	Sporadic inclusion body myositis: possible pathogenesis inferred from biomarkers. Current Opinion in Neurology, 2010, 23, 482-488.	3.6	47
33	Regulation of SQSTM1/p62 via UBA domain ubiquitination and its role in disease. Autophagy, 2017, 13, 1615-1616.	9.1	47
34	Myopathy-causing Mutations in an HSP40 Chaperone Disrupt Processing of Specific Client Conformers. Journal of Biological Chemistry, 2014, 289, 21120-21130.	3.4	46
35	VCP maintains lysosomal homeostasis and TFEB activity in differentiated skeletal muscle. Autophagy, 2019, 15, 1082-1099.	9.1	44
36	Quantification of autophagy flux using LC3 ELISA. Analytical Biochemistry, 2017, 530, 57-67.	2.4	43

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37	CANOMAD and other chronic ataxic neuropathies with disialosyl antibodies (CANDA). Journal of Neurology, 2018, 265, 1402-1409.	3.6	40
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	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
40	Increased autophagy accelerates colchicine-induced muscle toxicity. Autophagy, 2013, 9, 2115-2125.	9.1	35
41	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
42	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
43	Multisystem Proteinopathy Due to VCP Mutations: A Review of Clinical Heterogeneity and Genetic Diagnosis. Genes, 2022, 13, 963.	2.4	28
44	Neuronal VCP loss of function recapitulates FTLD-TDP pathology. Cell Reports, 2021, 36, 109399.	6.4	25
45	Mutations in the J domain of DNAJB6 cause dominant distal myopathy. Neuromuscular Disorders, 2020, 30, 38-46.	0.6	20
46	Autophagic vacuolar pathology in desminopathies. Neuromuscular Disorders, 2015, 25, 199-206.	0.6	19
47	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
48	Rare Manifestation of a c.290 C>T, p.Gly97GluVCPMutation. Case Reports in Genetics, 2015, 2015, 1-5.	0.2	16
49	A metastable subproteome underlies inclusion formation in muscle proteinopathies. Acta Neuropathologica Communications, 2019, 7, 197.	5.2	16
50	Novel GNE mutations in two phenotypically distinct HIBM2 patients. Neuromuscular Disorders, 2011, 21, 102-105.	0.6	15
51	Limbâ€girdle muscular dystrophy: A perspective from adult patients on what matters most. Muscle and Nerve, 2019, 60, 419-424.	2.2	15
52	Lithium chloride corrects weakness and myopathology in a preclinical model of LGMD1D. Neurology: Genetics, 2019, 5, e318.	1.9	15
53	VCP suppresses proteopathic seeding in neurons. Molecular Neurodegeneration, 2022, 17, 30.	10.8	15
54	Inhibition of DNAJ-HSP70 interaction improves strength in muscular dystrophy. Journal of Clinical Investigation, 2020, 130, 4470-4485.	8.2	14

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55	Monitoring Autophagy in the Treatment of Protein Aggregate Diseases: Steps Toward Identifying Autophagic Biomarkers. Neurotherapeutics, 2013, 10, 383-390.	4.4	11
56	Phenotypic diversity in an international Cure VCP Disease registry. Orphanet Journal of Rare Diseases, 2020, 15, 267.	2.7	11
57	Homozygous recessive MYH2 mutation mimicking dominant MYH2 associated myopathy. Neuromuscular Disorders, 2018, 28, 675-679.	0.6	10
58	Comparisons of ELISA and Western blot assays for detection of autophagy flux. Data in Brief, 2017, 13, 696-699.	1.0	8
59	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
60	Another VCP interactor: NF is enough. Journal of Clinical Investigation, 2011, 121, 4627-4630.	8.2	7
61	Client processing is altered by novel myopathy-causing mutations in the HSP40 J domain. PLoS ONE, 2020, 15, e0234207.	2.5	6
62	Sporadic Inclusion Body Myositis and Other Rimmed Vacuolar Myopathies. CONTINUUM Lifelong Learning in Neurology, 2019, 25, 1586-1598.	0.8	6
63	Cystinosis distal myopathy, novel clinical, pathological and genetic features. Neuromuscular Disorders, 2017, 27, 873-878.	0.6	5
64	Optimizing hand-function patient outcome measures for inclusion body myositis. Neuromuscular Disorders, 2020, 30, 807-814.	0.6	5
65	Loss-of-function mutation in VCP mimics the characteristic pathology as in FTLD-TARDBP. Autophagy, 2021, 17, 4502-4503.	9.1	5
66	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
67	Current and Future Approaches to Classify VUSs in LGMD-Related Genes. Genes, 2022, 13, 382.	2.4	3
68	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
69	Establishing prevalence in rare neuromuscular diseases. Neurology: Genetics, 2017, 3, e146.	1.9	1
70	Mdx mice have a defect in autophagy that is restored by rapamycinâ€loaded nanoparticle treatment. FASEB Journal, 2012, 26, 396.5.	0.5	1