Elena Miranda

List of Publications by Year
in descending order

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68 papers	4,145 citations	201575 27 h-index	114418 63 g-index
70 all docs	70 docs citations	70 times ranked	5168 citing authors

#	Article	IF	Citations
1	Targeted gene correction of Î±1-antitrypsin deficiency in induced pluripotent stem cells. Nature, 2011, 478, 391-394.	13.7	635
2	Modeling inherited metabolic disorders of the liver using human induced pluripotent stem cells. Journal of Clinical Investigation, 2010, 120, 3127-3136.	3.9	534
3	Endoplasmic reticulum dysfunction in neurological disease. Lancet Neurology, The, 2013, 12, 105-118.	4.9	396
4	Intraneuronal AÎ², non-amyloid aggregates and neurodegeneration in a Drosophila model of Alzheimerâ€™s disease. Neuroscience, 2005, 132, 123-135.	1.1	320
5	A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with Î±₁-antitrypsin deficiency. Hepatology, 2010, 52, 1078-1088.	3.6	138
6	Defining the mechanism of polymerization in the serpinopathies. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 17146-17151.	3.3	135
7	Neuroserpin: a serpin to think about. Cellular and Molecular Life Sciences, 2006, 63, 709-722.	2.4	125
8	Endoplasmic Reticulum-associated Degradation (ERAD) and Autophagy Cooperate to Degrade Polymerogenic Mutant Serpins. Journal of Biological Chemistry, 2009, 284, 22793-22802.	1.6	123
9	Endoplasmic reticulum polymers impair luminal protein mobility and sensitize to cellular stress in alpha₁â€antitrypsin deficiency. Hepatology, 2013, 57, 2049-2060.	3.6	108
10	Plasma and CSF serpins in Alzheimer disease and dementia with Lewy bodies. Neurology, 2007, 69, 1569-1579.	1.5	105
11	Mutants of Neuroserpin That Cause Dementia Accumulate as Polymers within the Endoplasmic Reticulum. Journal of Biological Chemistry, 2004, 279, 28283-28291.	1.6	102
12	The intracellular accumulation of polymeric neuroserpin explains the severity of the dementia FENIB. Human Molecular Genetics, 2008, 17, 1527-1539.	1.4	95
13	B-type Eph receptors and ephrins induce growth cone collapse through distinct intracellular pathways. Journal of Neurobiology, 2003, 57, 323-336.	3.7	86
14	ANCA-associated vasculitis is linked to carriage of the Z allele of Î±₁ antitrypsin and its polymers. Annals of the Rheumatic Diseases, 2011, 70, 1851-1856.	0.5	69
15	Circulating polymers in Â1-antitrypsin deficiency. European Respiratory Journal, 2014, 43, 1501-1504.	3.1	69
16	Neuroserpin Polymers Activate NF-ÎºB by a Calcium Signaling Pathway That Is Independent of the Unfolded Protein Response. Journal of Biological Chemistry, 2009, 284, 18202-18209.	1.6	68
17	Molecular mousetraps and the serpinopathies1. Biochemical Society Transactions, 2005, 33, 321-330.	1.6	59
18	Practical genetics: alpha-1-antitrypsin deficiency and the serpinopathies. European Journal of Human Genetics, 2004, 12, 167-172.	1.4	56

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19	Physiological modulation of BiP activity by trans-protomer engagement of the interdomain linker. ELife, 2015, 4, e08961.	2.8	55
20	Î±1-Antitrypsin deficiency, chronic obstructive pulmonary disease and the serpinopathies. Clinical Science, 2009, 116, 837-850.	1.8	51
21	A singleâ€chain variable fragment intrabody prevents intracellular polymerization of Z Î± ₁ â€antitrypsin while allowing its antiproteinase activity. FASEB Journal, 2015, 29, 2667-2678.	0.2	44
22	Three New Alpha1-Antitrypsin Deficiency Variants Help to Define a C-Terminal Region Regulating Conformational Change and Polymerization. PLoS ONE, 2012, 7, e38405.	1.1	43
23	Polymers of Z Î±₁-antitrypsin are secreted in cell models of disease. European Respiratory Journal, 2016, 47, 1005-1009.	3.1	41
24	The Serpinopathies. Methods in Enzymology, 2011, 501, 421-466.	0.4	35
25	Crystallographic and Cellular Characterisation of Two Mechanisms Stabilising the Native Fold of Î±1-Antitrypsin: Implications for Disease and Drug Design. Journal of Molecular Biology, 2009, 387, 857-868.	2.0	34
26	Characterisation of serpin polymers in vitro and in vivo. Methods, 2011, 53, 255-266.	1.9	31
27	Multiple roles of Activin/Nodal, bone morphogenetic protein, fibroblast growth factor and Wnt/Î²-catenin signalling in the anterior neural patterning of adherent human embryonic stem cell cultures. Open Biology, 2013, 3, 120167.	1.5	30
28	Molecular characterization of the new defective P_{brescia}alpha1-antitrypsin allele. Human Mutation, 2009, 30, E771-E781.	1.1	27
29	Characterising the association of latency with Î±1-antitrypsin polymerisation using a novel monoclonal antibody. International Journal of Biochemistry and Cell Biology, 2015, 58, 81-91.	1.2	26
30	The structural basis for Z Î± ₁ -antitrypsin polymerization in the liver. Science Advances, 2020, 6, .	4.7	26
31	Association between neuroserpin and molecular markers of brain damage in patients with acute ischemic stroke. Journal of Translational Medicine, 2011, 9, 58.	1.8	25
32	Neuroserpin polymers cause oxidative stress in a neuronal model of the dementia FENIB. Neurobiology of Disease, 2017, 103, 32-44.	2.1	25
33	The pathological Trento variant of alphaâ€1â€antitrypsin (E75V) shows nonclassical behaviour during polymerization. FEBS Journal, 2017, 284, 2110-2126.	2.2	23
34	The natural tissue plasminogen activator inhibitor neuroserpin and acute ischaemic stroke outcome. Thrombosis and Haemostasis, 2011, 105, 421-429.	1.8	22
35	An antibody raised against a pathogenic serpin variant induces mutant-like behaviour in the wild-type protein. Biochemical Journal, 2015, 468, 99-108.	1.7	22
36	Î±1-Antitrypsin Polymerizes in Alveolar Macrophages of Smokers With and Without Î±1-Antitrypsin Deficiency. Chest, 2018, 154, 607-616.	0.4	22

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37	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. Thrombosis and Haemostasis, 2007, 97, 394-399.	1.8	21
38	A Novel Interaction Between Aging and ER Overload in a Protein Conformational Dementia. Genetics, 2013, 193, 865-876.	1.2	21
39	Polymerisation underlies alpha1-antitrypsin deficiency, dementia and other serpinopathies. Frontiers in Bioscience - Landmark, 2004, 9, 2873.	3.0	19
40	Interactions between Nâ€linked glycosylation and polymerisation of neuroserpin within the endoplasmic reticulum. FEBS Journal, 2015, 282, 4565-4579.	2.2	19
41	Rostral floor plate (flexural organ) secretes glycoproteins immunologically similar to subcommissural organ glycoproteins in dogfish (Scyliorhinus canicula) embryos. Developmental Brain Research, 1997, 102, 69-75.	2.1	16
42	Neuroserpin: structure, function, physiology and pathology. Cellular and Molecular Life Sciences, 2021, 78, 6409-6430.	2.4	16
43	Intrahepatic heteropolymerization of M and Z alpha-1-antitrypsin. JCI Insight, 2020, 5, .	2.3	16
44	Targeting Serpins in High-Throughput and Structure-Based Drug Design. Methods in Enzymology, 2011, 501, 139-175.	0.4	15
45	An antibody that prevents serpin polymerisation acts by inducing a novel allosteric behaviour. Biochemical Journal, 2016, 473, 3269-3290.	1.7	15
46	Analysis and quantification of the secretory products of the subcommissural organ by use of monoclonal antibodies. Microscopy Research and Technique, 2001, 52, 510-519.	1.2	14
47	Quantification of the secretory glycoproteins of the subcommissural organ by a sensitive sandwich ELISA with a polyclonal antibody and a set of monoclonal antibodies against the bovine Reissner's fiber. Cell and Tissue Research, 1998, 294, 407-413.	1.5	13
48	Embelin binds to human neuroserpin and impairs its polymerisation. Scientific Reports, 2016, 6, 18769.	1.6	13
49	Association between circulating alpha-1 antitrypsin polymers and lung and liver disease. Respiratory Research, 2021, 22, 244.	1.4	13
50	Searching for specific binding sites of the secretory glycoproteins of the subcommissural organ. Microscopy Research and Technique, 2001, 52, 541-551.	1.2	12
51	Continuous delivery of a monoclonal antibody against Reissnerâ€™s fiber into CSF reveals CSF-soluble material immunorelated to the subcommissural organ in early chick embryos. Cell and Tissue Research, 2006, 326, 771-786.	1.5	11
52	Evaluation of Full-length, Cleaved and Nitrosylated Serum Surfactant Protein D as Biomarkers for COPD. COPD: Journal of Chronic Obstructive Pulmonary Disease, 2011, 8, 79-95.	0.7	11
53	The effects of weekly augmentation therapy in patients with PiZZ α1-antitrypsin deficiency. International Journal of COPD, 2012, 7, 687.	0.9	11
54	Glycosylation Tunes Neuroserpin Physiological and Pathological Properties. International Journal of Molecular Sciences, 2020, 21, 3235.	1.8	11

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55	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. Thrombosis and Haemostasis, 2007, 97, 394-9.	1.8	11
56	Embelin as Lead Compound for New Neuroserpin Polymerization Inhibitors. Life, 2020, 10, 111.	1.1	10
57	Cellular Models for the Serpinopathies. Methods in Molecular Biology, 2018, 1826, 109-121.	0.4	9
58	The molecular species responsible for Î± 1 â€antitrypsin deficiency are suppressed by a small molecule chaperone. FEBS Journal, 2021, 288, 2222-2237.	2.2	8
59	G392E neuroserpin causing the dementia FENIB is secreted from cells but is not synaptotoxic. Scientific Reports, 2021, 11, 8766.	1.6	7
60	The stability and activity of human neuroserpin are modulated by a salt bridge that stabilises the reactive centre loop. Scientific Reports, 2015, 5, 13666.	1.6	6
61	The Alpha-1 Antitrypsin Polymer Load Correlates With Hepatocyte Senescence, Fibrosis Stage and Liver-Related Mortality. Chronic Obstructive Pulmonary Diseases (Miami, Fla), 2020, 7, 151-162.	0.5	6
62	The Importance of N186 in the Alpha-1-Antitrypsin Shutter Region Is Revealed by the Novel Bologna Deficiency Variant. International Journal of Molecular Sciences, 2021, 22, 5668.	1.8	5
63	Serpin neuropathology in the P497S UBQLN2 mouse model of ALS/FTD. Brain Pathology, 2021, 31, e12948.	2.1	4
64	Functional analysis of novel alpha-1 antitrypsin variants G320R and V321F. Molecular Biology Reports, 2014, 41, 6133-6141.	1.0	3
65	Polymer toxicity in neurodegeneration FENIB. Oncotarget, 2017, 8, 35490-35491.	0.8	2
66	Neuroserpin Inclusion Bodies in a FENIB Yeast Model. Microorganisms, 2021, 9, 1498.	1.6	1
67	Role of cellular oxidative stress in dementia. , 2020, , 147-161.		1
68	Elucidating the pathological mechanisms of neurodegeneration in the lethal serpinopathy FENIB. Neural Regeneration Research, 2022, 17, 1733.	1.6	0