Elena Miranda

List of Publications by Year in Descending Order

66 papers	3,449 citations	26 h-index	58 g-index
70 ext. papers	3,841 ext. citations	6.6 avg, IF	4.59 L-index

#	Paper	IF	Citations
66	Elucidating the pathological mechanisms of neurodegeneration in the lethal serpinopathy FENIB.. *Neural Regeneration Research*, **2022**, 17, 1733-1734	4.5	
65	Serpin neuropathology in the P497S UBQLN2 mouse model of ALS/FTD. *Brain Pathology*, **2021**, 31, e12968	6	2
64	G392E neuroserpin causing the dementia FENIB is secreted from cells but is not synaptotoxic. *Scientific Reports*, **2021**, 11, 8766	4.9	4
63	Neuroserpin Inclusion Bodies in a FENIB Yeast Model. *Microorganisms*, **2021**, 9,	4.9	1
62	The molecular species responsible for α1-antitrypsin deficiency are suppressed by a small molecule chaperone. *FEBS Journal*, **2021**, 288, 2222-2237	5.7	2
61	Neuroserpin: structure, function, physiology and pathology. *Cellular and Molecular Life Sciences*, **2021**, 78, 6409-6430	10.3	2
60	Association between circulating alpha-1 antitrypsin polymers and lung and liver disease. *Respiratory Research*, **2021**, 22, 244	7.3	1
59	Glycosylation Tunes Neuroserpin Physiological and Pathological Properties. *International Journal of Molecular Sciences*, **2020**, 21,	6.3	9
58	Intrahepatic heteropolymerization of M and Z alpha-1-antitrypsin. *JCI Insight*, **2020**, 5,	9.9	5
57	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	2.7	3
56	Role of cellular oxidative stress in dementia **2020**, 147-161		
55	Embelin as Lead Compound for New Neuroserpin Polymerization Inhibitors. *Life*, **2020**, 10,	3	3
54	The structural basis for Z α1-antitrypsin polymerization in the liver. *Science Advances*, **2020**, 6,	14.3	12
53	Cellular Models for the Serpinopathies. *Methods in Molecular Biology*, **2018**, 1826, 109-121	1.4	3
52	α1-Antitrypsin Polymerizes in Alveolar Macrophages of Smokers With and Without α1-Antitrypsin Deficiency. *Chest*, **2018**, 154, 607-616	5.3	11
51	The pathological Trento variant of alpha-1-antitrypsin (E75V) shows nonclassical behaviour during polymerization. *FEBS Journal*, **2017**, 284, 2110-2126	5.7	16
50	Neuroserpin polymers cause oxidative stress in a neuronal model of the dementia FENIB. *Neurobiology of Disease*, **2017**, 103, 32-44	7.5	18

49	Polymers of Z 1-antitrypsin are secreted in cell models of disease. *European Respiratory Journal*, **2016**, 47, 1005-9	13.6	27
48	An antibody that prevents serpin polymerisation acts by inducing a novel allosteric behaviour. *Biochemical Journal*, **2016**, 473, 3269-90	3.8	15
47	Embelin binds to human neuroserpin and impairs its polymerisation. *Scientific Reports*, **2016**, 6, 18769	4.9	11
46	An antibody raised against a pathogenic serpin variant induces mutant-like behaviour in the wild-type protein. *Biochemical Journal*, **2015**, 468, 99-108	3.8	19
45	A single-chain variable fragment intrabody prevents intracellular polymerization of Z 1-antitrypsin while allowing its antiproteinase activity. *FASEB Journal*, **2015**, 29, 2667-78	0.9	38
44	The stability and activity of human neuroserpin are modulated by a salt bridge that stabilises the reactive centre loop. *Scientific Reports*, **2015**, 5, 13666	4.9	6
43	Physiological modulation of BiP activity by trans-protomer engagement of the interdomain linker. *ELife*, **2015**, 4, e08961	8.9	40
42	Interactions between N-linked glycosylation and polymerisation of neuroserpin within the endoplasmic reticulum. *FEBS Journal*, **2015**, 282, 4565-79	5.7	16
41	Characterising the association of latency with (1)-antitrypsin polymerisation using a novel monoclonal antibody. *International Journal of Biochemistry and Cell Biology*, **2015**, 58, 81-91	5.6	22
40	Functional analysis of novel alpha-1 antitrypsin variants G320R and V321F. *Molecular Biology Reports*, **2014**, 41, 6133-41	2.8	3
39	Circulating polymers in 1-antitrypsin deficiency. *European Respiratory Journal*, **2014**, 43, 1501-4	13.6	50
38	A novel interaction between aging and ER overload in a protein conformational dementia. *Genetics*, **2013**, 193, 865-76	4	20
37	Endoplasmic reticulum dysfunction in neurological disease. *Lancet Neurology, The*, **2013**, 12, 105-18	24.1	332
36	Multiple roles of Activin/Nodal, bone morphogenetic protein, fibroblast growth factor and Wnt/Ecatenin signalling in the anterior neural patterning of adherent human embryonic stem cell cultures. *Open Biology*, **2013**, 3, 120167	7	25
35	Endoplasmic reticulum polymers impair luminal protein mobility and sensitize to cellular stress in alpha1-antitrypsin deficiency. *Hepatology*, **2013**, 57, 2049-60	11.2	87
34	Three new alpha1-antitrypsin deficiency variants help to define a C-terminal region regulating conformational change and polymerization. *PLoS ONE*, **2012**, 7, e38405	3.7	33
33	The effects of weekly augmentation therapy in patients with PiZZ 1-antitrypsin deficiency. *International Journal of COPD*, **2012**, 7, 687-96	3	10
32	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	2	10

31	Targeted gene correction of 1-antitrypsin deficiency in induced pluripotent stem cells. *Nature*, **2011**, 478, 391-4	50.4	557
30	Characterisation of serpin polymers in vitro and in vivo. *Methods*, **2011**, 53, 255-66	4.6	28
29	The natural tissue plasminogen activator inhibitor neuroserpin and acute ischaemic stroke outcome. *Thrombosis and Haemostasis*, **2011**, 105, 421-9	7	19
28	Association between neuroserpin and molecular markers of brain damage in patients with acute ischemic stroke. *Journal of Translational Medicine*, **2011**, 9, 58	8.5	22
27	The serpinopathies studying serpin polymerization in vivo. *Methods in Enzymology*, **2011**, 501, 421-66	1.7	32
26	ANCA-associated vasculitis is linked to carriage of the Z allele of antitrypsin and its polymers. *Annals of the Rheumatic Diseases*, **2011**, 70, 1851-6	2.4	60
25	Targeting serpins in high-throughput and structure-based drug design. *Methods in Enzymology*, **2011**, 501, 139-75	1.7	15
24	Defining the mechanism of polymerization in the serpinopathies. *Proceedings of the National Academy of Sciences of the United States of America*, **2010**, 107, 17146-51	11.5	120
23	A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with alpha1-antitrypsin deficiency. *Hepatology*, **2010**, 52, 1078-88	11.2	111
22	Modeling inherited metabolic disorders of the liver using human induced pluripotent stem cells. *Journal of Clinical Investigation*, **2010**, 120, 3127-36	15.9	457
21	Neuroserpin polymers activate NF-kappaB by a calcium signaling pathway that is independent of the unfolded protein response. *Journal of Biological Chemistry*, **2009**, 284, 18202-9	5.4	57
20	Endoplasmic reticulum-associated degradation (ERAD) and autophagy cooperate to degrade polymerogenic mutant serpins. *Journal of Biological Chemistry*, **2009**, 284, 22793-802	5.4	104
19	Molecular characterization of the new defective P(brescia) alpha1-antitrypsin allele. *Human Mutation*, **2009**, 30, E771-81	4.7	20
18	Crystallographic and cellular characterisation of two mechanisms stabilising the native fold of alpha1-antitrypsin: implications for disease and drug design. *Journal of Molecular Biology*, **2009**, 387, 857-68	6.5	33
17	alpha1-Antitrypsin deficiency, chronic obstructive pulmonary disease and the serpinopathies. *Clinical Science*, **2009**, 116, 837-50	6.5	48
16	The intracellular accumulation of polymeric neuroserpin explains the severity of the dementia FENIB. *Human Molecular Genetics*, **2008**, 17, 1527-39	5.6	83
15	Plasma and CSF serpins in Alzheimer disease and dementia with Lewy bodies. *Neurology*, **2007**, 69, 1569-79	6.3	81
14	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. *Thrombosis and Haemostasis*, **2007**, 97, 394-399	7	20

13	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. *Thrombosis and Haemostasis*, **2007**, 97, 394-9	7	10
12	Neuroserpin: a serpin to think about. *Cellular and Molecular Life Sciences*, **2006**, 63, 709-22	10.3	106
11	Continuous delivery of a monoclonal antibody against Reissner\'s fiber into CSF reveals CSF-soluble material immunorelated to the subcommissural organ in early chick embryos. *Cell and Tissue Research*, **2006**, 326, 771-86	4.2	11
10	Intraneuronal Abeta, non-amyloid aggregates and neurodegeneration in a Drosophila model of Alzheimer\'s disease. *Neuroscience*, **2005**, 132, 123-35	3.9	285
9	Molecular mousetraps and the serpinopathies. *Biochemical Society Transactions*, **2005**, 33, 321-30	5.1	53
8	Polymerisation underlies alpha1-antitrypsin deficiency, dementia and other serpinopathies. *Frontiers in Bioscience - Landmark*, **2004**, 9, 2873-91	2.8	19
7	Mutants of neuroserpin that cause dementia accumulate as polymers within the endoplasmic reticulum. *Journal of Biological Chemistry*, **2004**, 279, 28283-91	5.4	84
6	Practical genetics: alpha-1-antitrypsin deficiency and the serpinopathies. *European Journal of Human Genetics*, **2004**, 12, 167-72	5.3	37
5	B-type Eph receptors and ephrins induce growth cone collapse through distinct intracellular pathways. *Journal of Neurobiology*, **2003**, 57, 323-36		72
4	Analysis and quantification of the secretory products of the subcommissural organ by use of monoclonal antibodies. *Microscopy Research and Technique*, **2001**, 52, 510-9	2.8	12
3	Searching for specific binding sites of the secretory glycoproteins of the subcommissural organ. *Microscopy Research and Technique*, **2001**, 52, 541-51	2.8	11
2	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, 407-13	4.2	11
1	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		13