

# Elena Miranda

## List of Publications by Citations

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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	Targeted gene correction of $\alpha$ -antitrypsin deficiency in induced pluripotent stem cells. <i>Nature</i> , <b>2011</b> , 478, 391-4	50.4	557
65	Modeling inherited metabolic disorders of the liver using human induced pluripotent stem cells. <i>Journal of Clinical Investigation</i> , <b>2010</b> , 120, 3127-36	15.9	457
64	Endoplasmic reticulum dysfunction in neurological disease. <i>Lancet Neurology</i> , <b>2013</b> , 12, 105-18	24.1	332
63	Intraneuronal Abeta, non-amyloid aggregates and neurodegeneration in a Drosophila model of Alzheimer's disease. <i>Neuroscience</i> , <b>2005</b> , 132, 123-35	3.9	285
62	Defining the mechanism of polymerization in the serpinopathies. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , <b>2010</b> , 107, 17146-51	11.5	120
61	A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with alpha1-antitrypsin deficiency. <i>Hepatology</i> , <b>2010</b> , 52, 1078-88	11.2	111
60	Neuroserpin: a serpin to think about. <i>Cellular and Molecular Life Sciences</i> , <b>2006</b> , 63, 709-22	10.3	106
59	Endoplasmic reticulum-associated degradation (ERAD) and autophagy cooperate to degrade polymerogenic mutant serpins. <i>Journal of Biological Chemistry</i> , <b>2009</b> , 284, 22793-802	5.4	104
58	Endoplasmic reticulum polymers impair luminal protein mobility and sensitize to cellular stress in alpha1-antitrypsin deficiency. <i>Hepatology</i> , <b>2013</b> , 57, 2049-60	11.2	87
57	Mutants of neuroserpin that cause dementia accumulate as polymers within the endoplasmic reticulum. <i>Journal of Biological Chemistry</i> , <b>2004</b> , 279, 28283-91	5.4	84
56	The intracellular accumulation of polymeric neuroserpin explains the severity of the dementia FENIB. <i>Human Molecular Genetics</i> , <b>2008</b> , 17, 1527-39	5.6	83
55	Plasma and CSF serpins in Alzheimer disease and dementia with Lewy bodies. <i>Neurology</i> , <b>2007</b> , 69, 1569-73	7.9	81
54	B-type Eph receptors and ephrins induce growth cone collapse through distinct intracellular pathways. <i>Journal of Neurobiology</i> , <b>2003</b> , 57, 323-36		72
53	ANCA-associated vasculitis is linked to carriage of the Z allele of $\alpha$ 1-antitrypsin and its polymers. <i>Annals of the Rheumatic Diseases</i> , <b>2011</b> , 70, 1851-6	2.4	60
52	Neuroserpin polymers activate NF-kappaB by a calcium signaling pathway that is independent of the unfolded protein response. <i>Journal of Biological Chemistry</i> , <b>2009</b> , 284, 18202-9	5.4	57
51	Molecular mousetraps and the serpinopathies. <i>Biochemical Society Transactions</i> , <b>2005</b> , 33, 321-30	5.1	53
50	Circulating polymers in $\alpha$ -antitrypsin deficiency. <i>European Respiratory Journal</i> , <b>2014</b> , 43, 1501-4	13.6	50

49	alpha1-Antitrypsin deficiency, chronic obstructive pulmonary disease and the serpinopathies. <i>Clinical Science</i> , <b>2009</b> , 116, 837-50	6.5	48
48	Physiological modulation of BiP activity by trans-protomer engagement of the interdomain linker. <i>ELife</i> , <b>2015</b> , 4, e08961	8.9	40
47	A single-chain variable fragment intrabody prevents intracellular polymerization of Z $\alpha$ 1-antitrypsin while allowing its antiprotease activity. <i>FASEB Journal</i> , <b>2015</b> , 29, 2667-78	0.9	38
46	Practical genetics: alpha-1-antitrypsin deficiency and the serpinopathies. <i>European Journal of Human Genetics</i> , <b>2004</b> , 12, 167-72	5.3	37
45	Three new alpha1-antitrypsin deficiency variants help to define a C-terminal region regulating conformational change and polymerization. <i>PLoS ONE</i> , <b>2012</b> , 7, e38405	3.7	33
44	Crystallographic and cellular characterisation of two mechanisms stabilising the native fold of alpha1-antitrypsin: implications for disease and drug design. <i>Journal of Molecular Biology</i> , <b>2009</b> , 387, 857-68	6.5	33
43	The serpinopathies studying serpin polymerization in vivo. <i>Methods in Enzymology</i> , <b>2011</b> , 501, 421-66	1.7	32
42	Characterisation of serpin polymers in vitro and in vivo. <i>Methods</i> , <b>2011</b> , 53, 255-66	4.6	28
41	Polymers of Z $\alpha$ 1-antitrypsin are secreted in cell models of disease. <i>European Respiratory Journal</i> , <b>2016</b> , 47, 1005-9	13.6	27
40	Multiple roles of Activin/Nodal, bone morphogenetic protein, fibroblast growth factor and Wnt/ $\beta$ -catenin signalling in the anterior neural patterning of adherent human embryonic stem cell cultures. <i>Open Biology</i> , <b>2013</b> , 3, 120167	7	25
39	Characterising the association of latency with $\alpha$ 1-antitrypsin polymerisation using a novel monoclonal antibody. <i>International Journal of Biochemistry and Cell Biology</i> , <b>2015</b> , 58, 81-91	5.6	22
38	Association between neuroserpin and molecular markers of brain damage in patients with acute ischemic stroke. <i>Journal of Translational Medicine</i> , <b>2011</b> , 9, 58	8.5	22
37	A novel interaction between aging and ER overload in a protein conformational dementia. <i>Genetics</i> , <b>2013</b> , 193, 865-76	4	20
36	Molecular characterization of the new defective P(brescia) alpha1-antitrypsin allele. <i>Human Mutation</i> , <b>2009</b> , 30, E771-81	4.7	20
35	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. <i>Thrombosis and Haemostasis</i> , <b>2007</b> , 97, 394-399	7	20
34	An antibody raised against a pathogenic serpin variant induces mutant-like behaviour in the wild-type protein. <i>Biochemical Journal</i> , <b>2015</b> , 468, 99-108	3.8	19
33	The natural tissue plasminogen activator inhibitor neuroserpin and acute ischaemic stroke outcome. <i>Thrombosis and Haemostasis</i> , <b>2011</b> , 105, 421-9	7	19
32	Polymerisation underlies alpha1-antitrypsin deficiency, dementia and other serpinopathies. <i>Frontiers in Bioscience - Landmark</i> , <b>2004</b> , 9, 2873-91	2.8	19

31	Neuroserpin polymers cause oxidative stress in a neuronal model of the dementia FENIB. <i>Neurobiology of Disease</i> , <b>2017</b> , 103, 32-44	7.5	18
30	The pathological Trento variant of alpha-1-antitrypsin (E75V) shows nonclassical behaviour during polymerization. <i>FEBS Journal</i> , <b>2017</b> , 284, 2110-2126	5.7	16
29	Interactions between N-linked glycosylation and polymerisation of neuroserpin within the endoplasmic reticulum. <i>FEBS Journal</i> , <b>2015</b> , 282, 4565-79	5.7	16
28	Targeting serpins in high-throughput and structure-based drug design. <i>Methods in Enzymology</i> , <b>2011</b> , 501, 139-75	1.7	15
27	An antibody that prevents serpin polymerisation acts by inducing a novel allosteric behaviour. <i>Biochemical Journal</i> , <b>2016</b> , 473, 3269-90	3.8	15
26	Rostral floor plate (flexural organ) secretes glycoproteins immunologically similar to subcommissural organ glycoproteins in dogfish ( <i>Scyliorhinus canicula</i> ) embryos. <i>Developmental Brain Research</i> , <b>1997</b> , 102, 69-75		13
25	Analysis and quantification of the secretory products of the subcommissural organ by use of monoclonal antibodies. <i>Microscopy Research and Technique</i> , <b>2001</b> , 52, 510-9	2.8	12
24	The structural basis for Z $\alpha$ 1-antitrypsin polymerization in the liver. <i>Science Advances</i> , <b>2020</b> , 6,	14.3	12
23	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. <i>Cell and Tissue Research</i> , <b>1998</b> , 294, 407-13	4.2	11
22	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. <i>Cell and Tissue Research</i> , <b>2006</b> , 326, 771-86	4.2	11
21	Searching for specific binding sites of the secretory glycoproteins of the subcommissural organ. <i>Microscopy Research and Technique</i> , <b>2001</b> , 52, 541-51	2.8	11
20	Embelin binds to human neuroserpin and impairs its polymerisation. <i>Scientific Reports</i> , <b>2016</b> , 6, 18769	4.9	11
19	$\alpha$ 1-Antitrypsin Polymerizes in Alveolar Macrophages of Smokers With and Without $\alpha$ 1-Antitrypsin Deficiency. <i>Chest</i> , <b>2018</b> , 154, 607-616	5.3	11
18	The effects of weekly augmentation therapy in patients with PiZZ $\alpha$ 1-antitrypsin deficiency. <i>International Journal of COPD</i> , <b>2012</b> , 7, 687-96	3	10
17	Evaluation of full-length, cleaved and nitrosylated serum surfactant protein D as biomarkers for COPD. <i>COPD: Journal of Chronic Obstructive Pulmonary Disease</i> , <b>2011</b> , 8, 79-95	2	10
16	Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. <i>Thrombosis and Haemostasis</i> , <b>2007</b> , 97, 394-9	7	10
15	Glycosylation Tunes Neuroserpin Physiological and Pathological Properties. <i>International Journal of Molecular Sciences</i> , <b>2020</b> , 21,	6.3	9
14	The stability and activity of human neuroserpin are modulated by a salt bridge that stabilises the reactive centre loop. <i>Scientific Reports</i> , <b>2015</b> , 5, 13666	4.9	6

13	Intrahepatic heteropolymerization of M and Z alpha-1-antitrypsin. <i>JCI Insight</i> , <b>2020</b> , 5,	9.9	5
12	G392E neuroserpin causing the dementia FENIB is secreted from cells but is not synaptotoxic. <i>Scientific Reports</i> , <b>2021</b> , 11, 8766	4.9	4
11	Functional analysis of novel alpha-1 antitrypsin variants G320R and V321F. <i>Molecular Biology Reports</i> , <b>2014</b> , 41, 6133-41	2.8	3
10	The Alpha-1 Antitrypsin Polymer Load Correlates With Hepatocyte Senescence, Fibrosis Stage and Liver-Related Mortality. <i>Chronic Obstructive Pulmonary Diseases (Miami, Fla)</i> , <b>2020</b> , 7, 151-162	2.7	3
9	Embelin as Lead Compound for New Neuroserpin Polymerization Inhibitors. <i>Life</i> , <b>2020</b> , 10,	3	3
8	Cellular Models for the Serpinopathies. <i>Methods in Molecular Biology</i> , <b>2018</b> , 1826, 109-121	1.4	3
7	Serpin neuropathology in the P497S UBQLN2 mouse model of ALS/FTD. <i>Brain Pathology</i> , <b>2021</b> , 31, e12948	1.8	2
6	The molecular species responsible for αantitrypsin deficiency are suppressed by a small molecule chaperone. <i>FEBS Journal</i> , <b>2021</b> , 288, 2222-2237	5.7	2
5	Neuroserpin: structure, function, physiology and pathology. <i>Cellular and Molecular Life Sciences</i> , <b>2021</b> , 78, 6409-6430	10.3	2
4	Neuroserpin Inclusion Bodies in a FENIB Yeast Model. <i>Microorganisms</i> , <b>2021</b> , 9,	4.9	1
3	Association between circulating alpha-1 antitrypsin polymers and lung and liver disease. <i>Respiratory Research</i> , <b>2021</b> , 22, 244	7.3	1
2	Elucidating the pathological mechanisms of neurodegeneration in the lethal serpinopathy FENIB.. <i>Neural Regeneration Research</i> , <b>2022</b> , 17, 1733-1734	4.5	
1	Role of cellular oxidative stress in dementia <b>2020</b> , 147-161		