

# Elena Miranda

## List of Publications by Year in descending order

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Version: 2024-02-01

68  
papers

4,145  
citations

201674

27  
h-index

114465

63  
g-index

70  
all docs

70  
docs citations

70  
times ranked

5168  
citing authors

| #  | ARTICLE   | IF   | CITATIONS |
|----|---|------|-----------|
| 1  | Elucidating the pathological mechanisms of neurodegeneration in the lethal serpinopathy FENIB. <i>Neural Regeneration Research</i> , 2022, 17, 1733.  | 3.0  | 0         |
| 2  | The molecular species responsible for $\alpha_1$ -antitrypsin deficiency are suppressed by a small molecule chaperone. <i>FEBS Journal</i> , 2021, 288, 2222-2237.                                      | 4.7  | 8         |
| 3  | Serpin neuropathology in the P497S UBQLN2 mouse model of ALS/FTD. <i>Brain Pathology</i> , 2021, 31, e12948.  | 4.1  | 4         |
| 4  | G392E neuroserpin causing the dementia FENIB is secreted from cells but is not synaptotoxic. <i>Scientific Reports</i> , 2021, 11, 8766.  | 3.3  | 7         |
| 5  | The Importance of N186 in the Alpha-1-Antitrypsin Shutter Region Is Revealed by the Novel Bologna Deficiency Variant. <i>International Journal of Molecular Sciences</i> , 2021, 22, 5668.              | 4.1  | 5         |
| 6  | Neuroserpin Inclusion Bodies in a FENIB Yeast Model. <i>Microorganisms</i> , 2021, 9, 1498.   | 3.6  | 1         |
| 7  | Neuroserpin: structure, function, physiology and pathology. <i>Cellular and Molecular Life Sciences</i> , 2021, 78, 6409-6430.  | 5.4  | 16        |
| 8  | Association between circulating alpha-1 antitrypsin polymers and lung and liver disease. <i>Respiratory Research</i> , 2021, 22, 244.   | 3.6  | 13        |
| 9  | Embelin as Lead Compound for New Neuroserpin Polymerization Inhibitors. <i>Life</i> , 2020, 10, 111.  | 2.4  | 10        |
| 10 | The structural basis for Z $\alpha_1$ -antitrypsin polymerization in the liver. <i>Science Advances</i> , 2020, 6, .  | 10.3 | 26        |
| 11 | Glycosylation Tunes Neuroserpin Physiological and Pathological Properties. <i>International Journal of Molecular Sciences</i> , 2020, 21, 3235.   | 4.1  | 11        |
| 12 | Intrahepatic heteropolymerization of M and Z alpha-1-antitrypsin. <i>JCI Insight</i> , 2020, 5, .   | 5.0  | 16        |
| 13 | The Alpha-1 Antitrypsin Polymer Load Correlates With Hepatocyte Senescence, Fibrosis Stage and Liver-Related Mortality. <i>Chronic Obstructive Pulmonary Diseases (Miami, Fla )</i> , 2020, 7, 151-162. | 0.7  | 6         |
| 14 | Role of cellular oxidative stress in dementia. , 2020, , 147-161.   |      | 1         |
| 15 | Cellular Models for the Serpinopathies. <i>Methods in Molecular Biology</i> , 2018, 1826, 109-121.  | 0.9  | 9         |
| 16 | $\alpha_1$ -Antitrypsin Polymerizes in Alveolar Macrophages of Smokers With and Without $\alpha_1$ -Antitrypsin Deficiency. <i>Chest</i> , 2018, 154, 607-616.  | 0.8  | 22        |
| 17 | The pathological Trento variant of alpha-1-antitrypsin (E75V) shows nonclassical behaviour during polymerization. <i>FEBS Journal</i> , 2017, 284, 2110-2126.   | 4.7  | 23        |
| 18 | Neuroserpin polymers cause oxidative stress in a neuronal model of the dementia FENIB. <i>Neurobiology of Disease</i> , 2017, 103, 32-44.   | 4.4  | 25        |

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|----|---|------|-----------|
| 19 | Polymer toxicity in neurodegeneration FENIB. <i>Oncotarget</i> , 2017, 8, 35490-35491.  | 1.8  | 2         |
| 20 | An antibody that prevents serpin polymerisation acts by inducing a novel allosteric behaviour. <i>Biochemical Journal</i> , 2016, 473, 3269-3290.   | 3.7  | 15        |
| 21 | Embelin binds to human neuroserpin and impairs its polymerisation. <i>Scientific Reports</i> , 2016, 6, 18769.  | 3.3  | 13        |
| 22 | Polymers of Z <sup>1</sup> -antitrypsin are secreted in cell models of disease. <i>European Respiratory Journal</i> , 2016, 47, 1005-1009.  | 6.7  | 41        |
| 23 | The stability and activity of human neuroserpin are modulated by a salt bridge that stabilises the reactive centre loop. <i>Scientific Reports</i> , 2015, 5, 13666.  | 3.3  | 6         |
| 24 | Physiological modulation of BiP activity by trans-protomer engagement of the interdomain linker. <i>ELife</i> , 2015, 4, e08961.  | 6.0  | 55        |
| 25 | Interactions between N-linked glycosylation and polymerisation of neuroserpin within the endoplasmic reticulum. <i>FEBS Journal</i> , 2015, 282, 4565-4579.   | 4.7  | 19        |
| 26 | Characterising the association of latency with Z <sup>1</sup> -antitrypsin polymerisation using a novel monoclonal antibody. <i>International Journal of Biochemistry and Cell Biology</i> , 2015, 58, 81-91.                                       | 2.8  | 26        |
| 27 | An antibody raised against a pathogenic serpin variant induces mutant-like behaviour in the wild-type protein. <i>Biochemical Journal</i> , 2015, 468, 99-108.  | 3.7  | 22        |
| 28 | A single-chain variable fragment intrabody prevents intracellular polymerization of Z <sup>1</sup> -antitrypsin while allowing its antiprotease activity. <i>FASEB Journal</i> , 2015, 29, 2667-2678.   | 0.5  | 44        |
| 29 | Circulating polymers in A <sup>1</sup> -antitrypsin deficiency. <i>European Respiratory Journal</i> , 2014, 43, 1501-1504.  | 6.7  | 69        |
| 30 | Functional analysis of novel alpha-1 antitrypsin variants G320R and V321F. <i>Molecular Biology Reports</i> , 2014, 41, 6133-6141.  | 2.3  | 3         |
| 31 | A Novel Interaction Between Aging and ER Overload in a Protein Conformational Dementia. <i>Genetics</i> , 2013, 193, 865-876.   | 2.9  | 21        |
| 32 | Endoplasmic reticulum dysfunction in neurological disease. <i>Lancet Neurology</i> , The, 2013, 12, 105-118.  | 10.2 | 396       |
| 33 | 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> , 2013, 3, 120167. | 3.6  | 30        |
| 34 | Endoplasmic reticulum polymers impair luminal protein mobility and sensitize to cellular stress in alpha <sup>1</sup> -antitrypsin deficiency. <i>Hepatology</i> , 2013, 57, 2049-2060.   | 7.3  | 108       |
| 35 | Three New Alpha1-Antitrypsin Deficiency Variants Help to Define a C-Terminal Region Regulating Conformational Change and Polymerization. <i>PLoS ONE</i> , 2012, 7, e38405.   | 2.5  | 43        |
| 36 | The effects of weekly augmentation therapy in patients with PiZZ &alpha;1-antitrypsin deficiency. <i>International Journal of COPD</i> , 2012, 7, 687.  | 2.3  | 11        |

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|----|---|------|-----------|
| 37 | 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.                                      | 1.6  | 11        |
| 38 | Targeted gene correction of $\alpha$ 1-antitrypsin deficiency in induced pluripotent stem cells. Nature, 2011, 478, 391-394.  | 27.8 | 635       |
| 39 | Characterisation of serpin polymers in vitro and in vivo. Methods, 2011, 53, 255-266.   | 3.8  | 31        |
| 40 | The natural tissue plasminogen activator inhibitor neuroserpin and acute ischaemic stroke outcome. Thrombosis and Haemostasis, 2011, 105, 421-429.  | 3.4  | 22        |
| 41 | Association between neuroserpin and molecular markers of brain damage in patients with acute ischemic stroke. Journal of Translational Medicine, 2011, 9, 58.   | 4.4  | 25        |
| 42 | The Serpinopathies. Methods in Enzymology, 2011, 501, 421-466.  | 1.0  | 35        |
| 43 | ANCA-associated vasculitis is linked to carriage of the Z allele of $\alpha$ 1-antitrypsin and its polymers. Annals of the Rheumatic Diseases, 2011, 70, 1851-1856.   | 0.9  | 69        |
| 44 | Targeting Serpins in High-Throughput and Structure-Based Drug Design. Methods in Enzymology, 2011, 501, 139-175.  | 1.0  | 15        |
| 45 | A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with $\alpha$ 1-antitrypsin deficiency. Hepatology, 2010, 52, 1078-1088.  | 7.3  | 138       |
| 46 | 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.  | 7.1  | 135       |
| 47 | Modeling inherited metabolic disorders of the liver using human induced pluripotent stem cells. Journal of Clinical Investigation, 2010, 120, 3127-3136.  | 8.2  | 534       |
| 48 | Neuroserpin Polymers Activate NF- $\kappa$ B by a Calcium Signaling Pathway That Is Independent of the Unfolded Protein Response. Journal of Biological Chemistry, 2009, 284, 18202-18209.                          | 3.4  | 68        |
| 49 | Endoplasmic Reticulum-associated Degradation (ERAD) and Autophagy Cooperate to Degrade Polymerogenic Mutant Serpins. Journal of Biological Chemistry, 2009, 284, 22793-22802.                                       | 3.4  | 123       |
| 50 | Molecular characterization of the new defective P <sup>brescia</sup> $\alpha$ 1-antitrypsin allele. Human Mutation, 2009, 30, E771-E781.  | 2.5  | 27        |
| 51 | Crystallographic and Cellular Characterisation of Two Mechanisms Stabilising the Native Fold of $\alpha$ 1-Antitrypsin: Implications for Disease and Drug Design. Journal of Molecular Biology, 2009, 387, 857-868. | 4.2  | 34        |
| 52 | $\alpha$ 1-Antitrypsin deficiency, chronic obstructive pulmonary disease and the serpinopathies. Clinical Science, 2009, 116, 837-850.  | 4.3  | 51        |
| 53 | The intracellular accumulation of polymeric neuroserpin explains the severity of the dementia FENIB. Human Molecular Genetics, 2008, 17, 1527-1539.   | 2.9  | 95        |
| 54 | Plasma and CSF serpins in Alzheimer disease and dementia with Lewy bodies. Neurology, 2007, 69, 1569-1579.  | 1.1  | 105       |

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|----|--|-----|-----------|
| 55 | Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. <i>Thrombosis and Haemostasis</i> , 2007, 97, 394-399.  | 3.4 | 21        |
| 56 | Expression of the serine protease inhibitor neuroserpin in cells of the human myeloid lineage. <i>Thrombosis and Haemostasis</i> , 2007, 97, 394-9.  | 3.4 | 11        |
| 57 | 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> , 2006, 326, 771-786.                              | 2.9 | 11        |
| 58 | Neuroserpin: a serpin to think about. <i>Cellular and Molecular Life Sciences</i> , 2006, 63, 709-722.   | 5.4 | 125       |
| 59 | Molecular mousetraps and the serpinopathies <sup>1</sup> . <i>Biochemical Society Transactions</i> , 2005, 33, 321-330.  | 3.4 | 59        |
| 60 | Intraneuronal A $\beta$ <sup>2</sup> , non-amyloid aggregates and neurodegeneration in a <i>Drosophila</i> model of Alzheimer's disease. <i>Neuroscience</i> , 2005, 132, 123-135.   | 2.3 | 320       |
| 61 | Polymerisation underlies alpha1-antitrypsin deficiency, dementia and other serpinopathies. <i>Frontiers in Bioscience - Landmark</i> , 2004, 9, 2873.  | 3.0 | 19        |
| 62 | Mutants of Neuroserpin That Cause Dementia Accumulate as Polymers within the Endoplasmic Reticulum. <i>Journal of Biological Chemistry</i> , 2004, 279, 28283-28291.   | 3.4 | 102       |
| 63 | Practical genetics: alpha-1-antitrypsin deficiency and the serpinopathies. <i>European Journal of Human Genetics</i> , 2004, 12, 167-172.  | 2.8 | 56        |
| 64 | B-type Eph receptors and ephrins induce growth cone collapse through distinct intracellular pathways. <i>Journal of Neurobiology</i> , 2003, 57, 323-336.  | 3.6 | 86        |
| 65 | Analysis and quantification of the secretory products of the subcommissural organ by use of monoclonal antibodies. <i>Microscopy Research and Technique</i> , 2001, 52, 510-519.   | 2.2 | 14        |
| 66 | Searching for specific binding sites of the secretory glycoproteins of the subcommissural organ. <i>Microscopy Research and Technique</i> , 2001, 52, 541-551.   | 2.2 | 12        |
| 67 | 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> , 1998, 294, 407-413. | 2.9 | 13        |
| 68 | 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> , 1997, 102, 69-75.                           | 1.7 | 16        |