

Angel L. Pey

List of Publications by Year in descending order

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87
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2,737
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172386

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214721

47
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docs citations

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times ranked

2763
citing authors

#	ARTICLE	IF	CITATIONS
1	Predicted Effects of Missense Mutations on Native-State Stability Account for Phenotypic Outcome in Phenylketonuria, a Paradigm of Misfolding Diseases. <i>American Journal of Human Genetics</i> , 2007, 81, 1006-1024.	2.6	157
2	From The Cover: Correction of kinetic and stability defects by tetrahydrobiopterin in phenylketonuria patients with certain phenylalanine hydroxylase mutations. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2004, 101, 16903-16908.	3.3	156
3	Identification of pharmacological chaperones as potential therapeutic agents to treat phenylketonuria. <i>Journal of Clinical Investigation</i> , 2008, 118, 2858-2867.	3.9	145
4	Primary hyperoxalurias: Disorders of glyoxylate detoxification. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2012, 1822, 1453-1464.	1.8	124
5	Phenylketonuria: Genotype-phenotype correlations based on expression analysis of structural and functional mutations in PAH. <i>Human Mutation</i> , 2003, 21, 370-378.	1.1	111
6	Mechanisms underlying responsiveness to tetrahydrobiopterin in mild phenylketonuria mutations. <i>Human Mutation</i> , 2004, 24, 388-399.	1.1	109
7	Human cystathionine β -synthase (CBS) contains two classes of binding sites for S-adenosylmethionine (SAM): complex regulation of CBS activity and stability by SAM. <i>Biochemical Journal</i> , 2013, 449, 109-121.	1.7	78
8	NQO1: A target for the treatment of cancer and neurological diseases, and a model to understand loss of function disease mechanisms. <i>Biochimica Et Biophysica Acta - Proteins and Proteomics</i> , 2019, 1867, 663-676.	1.1	68
9	Structure of Phenylalanine Hydroxylase from <i>Colwellia psychroerythraea</i> 34H, a Monomeric Cold Active Enzyme with Local Flexibility around the Active Site and High Overall Stability. <i>Journal of Biological Chemistry</i> , 2007, 282, 21973-21986.	1.6	62
10	Effects of prolonged stanozolol treatment on antioxidant enzyme activities, oxidative stress markers, and heat shock protein HSP72 levels in rat liver. <i>Journal of Steroid Biochemistry and Molecular Biology</i> , 2003, 87, 269-277.	1.2	56
11	FAD binding overcomes defects in activity and stability displayed by cancer-associated variants of human NQO1. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2014, 1842, 2163-2173.	1.8	56
12	NAD(P)H quinone oxidoreductase (NQO1): an enzyme which needs just enough mobility, in just the right places. <i>Bioscience Reports</i> , 2019, 39, .	1.1	55
13	Role of low native state kinetic stability and interaction of partially unfolded states with molecular chaperones in the mitochondrial protein mistargeting associated with primary hyperoxaluria. <i>Amino Acids</i> , 2011, 41, 1233-1245.	1.2	54
14	Structural Basis of the Oncogenic Interaction of Phosphatase PRL-1 with the Magnesium Transporter CNNM2. <i>Journal of Biological Chemistry</i> , 2017, 292, 786-801.	1.6	48
15	The Role of Protein Denaturation Energetics and Molecular Chaperones in the Aggregation and Mistargeting of Mutants Causing Primary Hyperoxaluria Type I. <i>PLoS ONE</i> , 2013, 8, e71963.	1.1	48
16	Molecular basis of classic galactosemia from the structure of human galactose 1-phosphate uridylyltransferase. <i>Human Molecular Genetics</i> , 2016, 25, 2234-2244.	1.4	43
17	Domain Organization, Catalysis and Regulation of Eukaryotic Cystathionine Beta-Synthases. <i>PLoS ONE</i> , 2014, 9, e105290.	1.1	42
18	Protein Homeostasis Defects of Alanine-Glyoxylate Aminotransferase: New Therapeutic Strategies in Primary Hyperoxaluria Type I. <i>BioMed Research International</i> , 2013, 2013, 1-15.	0.9	40

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19	Engineering proteins with tunable thermodynamic and kinetic stabilities. <i>Proteins: Structure, Function and Bioinformatics</i> , 2008, 71, 165-174.	1.5	39
20	Conformational dynamics is key to understanding loss-of-function of NQO1 cancer-associated polymorphisms and its correction by pharmacological ligands. <i>Scientific Reports</i> , 2016, 6, 20331.	1.6	39
21	Intrinsically disordered chromatin protein NUPR1 binds to the C-terminal region of Polycomb RING1B. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2017, 114, E6332-E6341.	3.3	39
22	Specific interaction of the diastereomers 7(R)- and 7(S)-tetrahydrobiopterin with phenylalanine hydroxylase: implications for understanding primapterinuria and vitiligo. <i>FASEB Journal</i> , 2006, 20, 2130-2132.	0.2	37
23	Thermodynamic Characterization of the Binding of Tetrahydropterins to Phenylalanine Hydroxylase. <i>Journal of the American Chemical Society</i> , 2004, 126, 13670-13678.	6.6	36
24	Site-to-site interdomain communication may mediate different loss-of-function mechanisms in a cancer-associated NQO1 polymorphism. <i>Scientific Reports</i> , 2017, 7, 44532.	1.6	35
25	Enhanced vulnerability of human proteins towards disease-associated inactivation through divergent evolution. <i>Human Molecular Genetics</i> , 2017, 26, 3531-3544.	1.4	34
26	Second Generation of Mannich Base-Type Derivatives with <i>in Vivo</i> Activity against <i>Trypanosoma cruzi</i> . <i>Journal of Medicinal Chemistry</i> , 2018, 61, 5643-5663.	2.9	32
27	Rescuing Proteins of Low Kinetic Stability by Chaperones and Natural Ligands: Phenylketonuria, a Case Study. <i>Progress in Molecular Biology and Translational Science</i> , 2008, 83, 89-134.	0.9	31
28	Modulation of Buried Ionizable Groups in Proteins with Engineered Surface Charge. <i>Journal of the American Chemical Society</i> , 2010, 132, 1218-1219.	6.6	31
29	Natural Small Molecules as Stabilizers and Activators of Cancer-Associated NQO1 Polymorphisms. <i>Current Drug Targets</i> , 2016, 17, 1506-1514.	1.0	31
30	The activity of wild-type and mutant phenylalanine hydroxylase and its regulation by phenylalanine and tetrahydrobiopterin at physiological and pathological concentrations: An isothermal titration calorimetry study. <i>Molecular Genetics and Metabolism</i> , 2005, 86, 43-53.	0.5	30
31	The consensus-based approach for gene/enzyme replacement therapies and crystallization strategies: the case of human alanine-glyoxylate aminotransferase. <i>Biochemical Journal</i> , 2014, 462, 453-463.	1.7	30
32	Targeting Cystathionine Beta-Synthase Misfolding in Homocystinuria by Small Ligands: State of the Art and Future Directions. <i>Current Drug Targets</i> , 2016, 17, 1455-1470.	1.0	30
33	Effect of pharmacological chaperones on brain tyrosine hydroxylase and tryptophan hydroxylase 2. <i>Journal of Neurochemistry</i> , 2010, 114, 853-863.	2.1	29
34	Insight into the specificity and severity of pathogenic mechanisms associated with missense mutations through experimental and structural perturbation analyses. <i>Human Molecular Genetics</i> , 2019, 28, 1-15.	1.4	29
35	Potential Pharmacological Chaperones for Cystathionine Beta-Synthase-Deficient Homocystinuria. <i>Handbook of Experimental Pharmacology</i> , 2017, 245, 345-383.	0.9	28
36	BANK1 interacts with TRAF6 and MyD88 in innate immune signaling in B cells. <i>Cellular and Molecular Immunology</i> , 2020, 17, 954-965.	4.8	28

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37	Anabolic function of phenylalanine hydroxylase in <i>Caenorhabditis elegans</i> . <i>FASEB Journal</i> , 2008, 22, 3046-3058.	0.2	27
38	Biochemical characterization of mutant phenylalanine hydroxylase enzymes and correlation with clinical presentation in hyperphenylalaninaemic patients. <i>Journal of Inherited Metabolic Disease</i> , 2009, 32, 10-21.	1.7	27
39	Structural and Energetic Basis of Protein Kinetic Destabilization in Human Phosphoglycerate Kinase 1 Deficiency. <i>Biochemistry</i> , 2013, 52, 1160-1170.	1.2	26
40	Thermodynamics of cooperative binding of FAD to human NQO1: Implications to understanding cofactor-dependent function and stability of the flavoproteome. <i>Archives of Biochemistry and Biophysics</i> , 2017, 636, 17-27.	1.4	26
41	The metastability of human UDP-galactose 4-epimerase (GALE) is increased by variants associated with type III galactosemia but decreased by substrate and cofactor binding. <i>Archives of Biochemistry and Biophysics</i> , 2014, 562, 103-114.	1.4	25
42	Conformational Properties of Nine Purified Cystathionine β -Synthase Mutants. <i>Biochemistry</i> , 2012, 51, 4755-4763.	1.2	24
43	Kinetic stability of cystathionine beta-synthase can be modulated by structural analogs of S-adenosylmethionine: Potential approach to pharmacological chaperone therapy for homocystinuria. <i>Biochimie</i> , 2016, 126, 6-13.	1.3	23
44	Insights into human phosphoglycerate kinase 1 deficiency as a conformational disease from biochemical, biophysical, and in vitro expression analyses. <i>Journal of Inherited Metabolic Disease</i> , 2014, 37, 909-916.	1.7	22
45	The Catalytic Cycle of the Antioxidant and Cancer-Associated Human NQO1 Enzyme: Hydride Transfer, Conformational Dynamics and Functional Cooperativity. <i>Antioxidants</i> , 2020, 9, 772.	2.2	22
46	Structural basis of the pleiotropic and specific phenotypic consequences of missense mutations in the multifunctional NAD(P)H:quinone oxidoreductase 1 and their pharmacological rescue. <i>Redox Biology</i> , 2021, 46, 102112.	3.9	22
47	Protein homeostasis disorders of key enzymes of amino acids metabolism: mutation-induced protein kinetic destabilization and new therapeutic strategies. <i>Amino Acids</i> , 2013, 45, 1331-1341.	1.2	21
48	A mechanism for cancer-associated inactivation of NQO1 due to P187S and its reactivation by the consensus mutation H80R. <i>FEBS Letters</i> , 2017, 591, 2826-2835.	1.3	21
49	Biophysical and functional perturbation analyses at cancer-associated P187 and K240 sites of the multifunctional NAD(P)H:quinone oxidoreductase 1. <i>International Journal of Biological Macromolecules</i> , 2018, 118, 1912-1923.	3.6	21
50	A Dynamic Core in Human NQO1 Controls the Functional and Stability Effects of Ligand Binding and Their Communication across the Enzyme Dimer. <i>Biomolecules</i> , 2019, 9, 728.	1.8	21
51	Divergence in enzyme regulation between <i>Caenorhabditis elegans</i> and human tyrosine hydroxylase, the key enzyme in the synthesis of dopamine. <i>Biochemical Journal</i> , 2011, 434, 133-141.	1.7	20
52	The lower limits for protein stability and foldability in primary hyperoxaluria type I. <i>Biochimica Et Biophysica Acta - Proteins and Proteomics</i> , 2014, 1844, 2355-2365.	1.1	20
53	Iron binding effects on the kinetic stability and unfolding energetics of a thermophilic phenylalanine hydroxylase from <i>Chloroflexus aurantiacus</i> . <i>Journal of Biological Inorganic Chemistry</i> , 2009, 14, 521-531.	1.1	15
54	Protein Stability, Folding and Misfolding in Human PGK1 Deficiency. <i>Biomolecules</i> , 2013, 3, 1030-1052.	1.8	15

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55	Phosphorylation compromises FAD binding and intracellular stability of wild-type and cancer-associated NQO1: Insights into flavo-proteome stability. <i>International Journal of Biological Macromolecules</i> , 2019, 125, 1275-1288.	3.6	15
56	<i>Thermoplasma acidophilum</i> Cdc6 protein stimulates MCM helicase activity by regulating its ATPase activity. <i>Nucleic Acids Research</i> , 2008, 36, 5602-5609.	6.5	14
57	Superstoichiometric binding of L-Phe to phenylalanine hydroxylase from <i>Caenorhabditis elegans</i> : evolutionary implications. <i>Amino Acids</i> , 2010, 39, 1463-1475.	1.2	14
58	Molecular Recognition of PTS-1 Cargo Proteins by Pex5p: Implications for Protein Mistargeting in Primary Hyperoxaluria. <i>Biomolecules</i> , 2015, 5, 121-141.	1.8	14
59	Galactokinase promiscuity: a question of flexibility?. <i>Biochemical Society Transactions</i> , 2016, 44, 116-122.	1.6	14
60	Structural and functional insights on the roles of molecular chaperones in the mistargeting and aggregation phenotypes associated with primary hyperoxaluria type I. <i>Advances in Protein Chemistry and Structural Biology</i> , 2019, 114, 119-152.	1.0	14
61	The interplay between protein stability and dynamics in conformational diseases: The case of hPGK1 deficiency. <i>Biochimica Et Biophysica Acta - Proteins and Proteomics</i> , 2013, 1834, 2502-2511.	1.1	12
62	Evolutionary Divergent Suppressor Mutations in Conformational Diseases. <i>Genes</i> , 2018, 9, 352.	1.0	12
63	Warfarin increases thermal resistance of albumin through stabilization of the protein lobe that includes its binding site. <i>Archives of Biochemistry and Biophysics</i> , 2019, 676, 108123.	1.4	12
64	Systemic Alanine Glyoxylate Aminotransferase mRNA Improves Glyoxylate Metabolism in a Mouse Model of Primary Hyperoxaluria Type 1. <i>Nucleic Acid Therapeutics</i> , 2019, 29, 104-113.	2.0	12
65	Galactosemia: Towards Pharmacological Chaperones. <i>Journal of Personalized Medicine</i> , 2021, 11, 106.	1.1	12
66	Targeting HIF-1 α Function in Cancer through the Chaperone Action of NQO1: Implications of Genetic Diversity of NQO1. <i>Journal of Personalized Medicine</i> , 2022, 12, 747.	1.1	12
67	Allosteric Communication in the Multifunctional and Redox NQO1 Protein Studied by Cavity-Making Mutations. <i>Antioxidants</i> , 2022, 11, 1110.	2.2	12
68	Tetrahydrobiopterin for patients with phenylketonuria. <i>Lancet, The</i> , 2007, 370, 462-463.	6.3	11
69	The Regulatory Subunit of PKA-I Remains Partially Structured and Undergoes β -Aggregation upon Thermal Denaturation. <i>PLoS ONE</i> , 2011, 6, e17602.	1.1	11
70	The role of surface electrostatics on the stability, function and regulation of human cystathionine β -synthase, a complex multidomain and oligomeric protein. <i>Biochimica Et Biophysica Acta - Proteins and Proteomics</i> , 2014, 1844, 1453-1462.	1.1	10
71	Natural (and Unnatural) Small Molecules as Pharmacological Chaperones and Inhibitors in Cancer. <i>Handbook of Experimental Pharmacology</i> , 2017, 245, 155-190.	0.9	10
72	The chondroitin sulfate/dermatan sulfate 4-O-endosulfatase from marine bacterium <i>Vibrio</i> sp FC509 is a dimeric species: Biophysical characterization of an endosulfatase. <i>Biochimie</i> , 2016, 131, 85-95.	1.3	9

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73	Oligomeric status of human cystathionine beta-synthase modulates AdoMet binding. FEBS Letters, 2016, 590, 4461-4471.	1.3	8
74	Experimental and computational evidence on conformational fluctuations as a source of catalytic defects in genetic diseases. RSC Advances, 2016, 6, 58604-58612.	1.7	8
75	Improving the Activity and Stability of Human Galactokinase for Therapeutic and Biotechnological Applications. ChemBioChem, 2018, 19, 1088-1095.	1.3	8
76	Anion-specific interaction with human NQO1 inhibits flavin binding. International Journal of Biological Macromolecules, 2019, 126, 1223-1233.	3.6	8
77	Naturally-Occurring Rare Mutations Cause Mild to Catastrophic Effects in the Multifunctional and Cancer-Associated NQO1 Protein. Journal of Personalized Medicine, 2020, 10, 207.	1.1	8
78	New salicylic acid derivatives, double inhibitors of glycolate oxidase and lactate dehydrogenase, as effective agents decreasing oxalate production. European Journal of Medicinal Chemistry, 2022, 237, 114396.	2.6	7
79	Phenylalanine hydroxylase expression in primary rat hepatocytes is modulated by oxygen concentration. Molecular Genetics and Metabolism, 2010, 101, 279-281.	0.5	5
80	pH-dependent relationship between thermodynamic and kinetic stability in the denaturation of human phosphoglycerate kinase 1. Biochimie, 2014, 103, 7-15.	1.3	5
81	Dimerization Drives Proper Folding of Human Alanine:Glyoxylate Aminotransferase But Is Dispensable for Peroxisomal Targeting. Journal of Personalized Medicine, 2021, 11, 273.	1.1	5
82	Towards Accurate Genotype-Phenotype Correlations in the CYP2D6 Gene. Journal of Personalized Medicine, 2020, 10, 158.	1.1	4
83	Intrinsically disordered protein NUPR1 binds to the armadillo-repeat domain of Plakophilin 1. International Journal of Biological Macromolecules, 2021, 170, 549-560.	3.6	4
84	Caenorhabditis elegans AGXT-1 is a mitochondrial and temperature-adapted ortholog of peroxisomal human AGT1: New insights into between-species divergence in glyoxylate metabolism. Biochimica Et Biophysica Acta - Proteins and Proteomics, 2016, 1864, 1195-1205.	1.1	3
85	Protein homeostasis and disease. , 2020, , 23-37.		2
86	A single evolutionarily divergent mutation determines the different FAD-binding affinities of human and rat NQO1 due to site-specific phosphorylation. FEBS Letters, 2021, 596, 29.	1.3	2
87	Editorial (Thematic Issue: Targeting Conformational Diseases with Natural and Pharmacological) Tj ETQq1 1 0.784314 rgBT /Qverlock 10		