

Vicente Rubio

List of Publications by Year in descending order

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145
papers

4,891
citations

109321

35
h-index

118850

62
g-index

149
all docs

149
docs citations

149
times ranked

4501
citing authors

#	ARTICLE	IF	CITATIONS
1	Suggested guidelines for the diagnosis and management of urea cycle disorders. <i>Orphanet Journal of Rare Diseases</i> , 2012, 7, 32.	2.7	596
2	Structural Insight into Partner Specificity and Phosphoryl Transfer in Two-Component Signal Transduction. <i>Cell</i> , 2009, 139, 325-336.	28.9	351
3	Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision. <i>Journal of Inherited Metabolic Disease</i> , 2019, 42, 1192-1230.	3.6	277
4	The mechanism of signal transduction by two-component systems. <i>Current Opinion in Structural Biology</i> , 2010, 20, 763-771.	5.7	206
5	Structure of Acetylglutamate Kinase, a Key Enzyme for Arginine Biosynthesis and a Prototype for the Amino Acid Kinase Enzyme Family, during Catalysis. <i>Structure</i> , 2002, 10, 329-342.	3.3	126
6	Structural basis for the regulation of NtcA-dependent transcription by proteins PipX and PII. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2010, 107, 15397-15402.	7.1	116
7	The crystal structure of the complex of PII and acetylglutamate kinase reveals how PII controls the storage of nitrogen as arginine. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2007, 104, 17644-17649.	7.1	113
8	Gene Structure, Organization, Expression, and Potential Regulatory Mechanisms of Arginine Catabolism in <i>Enterococcus faecalis</i> . <i>Journal of Bacteriology</i> , 2002, 184, 6289-6300.	2.2	92
9	Arginine and nitrogen storage. <i>Current Opinion in Structural Biology</i> , 2008, 18, 673-681.	5.7	92
10	Structural and Functional Insights into Endoglin Ligand Recognition and Binding. <i>PLoS ONE</i> , 2012, 7, e29948.	2.5	86
11	Molecular defects in human carbamoyl phosphate synthetase I: mutational spectrum, diagnostic and protein structure considerations. <i>Human Mutation</i> , 2011, 32, 579-589.	2.5	67
12	Structure of human carbamoyl phosphate synthetase: deciphering the on/off switch of human ureagenesis. <i>Scientific Reports</i> , 2015, 5, 16950.	3.3	64
13	Structural Bases of Feed-back Control of Arginine Biosynthesis, Revealed by the Structures of Two Hexameric N-Acetylglutamate Kinases, from <i>Thermotoga maritima</i> and <i>Pseudomonas aeruginosa</i> . <i>Journal of Molecular Biology</i> , 2006, 356, 695-713.	4.2	63
14	A Novel Two-domain Architecture Within the Amino Acid Kinase Enzyme Family Revealed by the Crystal Structure of <i>Escherichia coli</i> Glutamate 5-kinase. <i>Journal of Molecular Biology</i> , 2007, 367, 1431-1446.	4.2	62
15	Domain structure of the large subunit of <i>Escherichia coli</i> carbamoyl phosphate synthetase. Location of the binding site for the allosteric inhibitor UMP in the carboxy-terminal domain. <i>Biochemistry</i> , 1991, 30, 1068-1075.	2.5	59
16	The Gene Cluster for Agmatine Catabolism of <i>Enterococcus faecalis</i> : Study of Recombinant Putrescine Transcarbamylase and Agmatine Deiminase and a Snapshot of Agmatine Deiminase Catalyzing Its Reaction. <i>Journal of Bacteriology</i> , 2007, 189, 1254-1265.	2.2	59
17	Improved cross-linked enzyme aggregates for the production of desacetyl β -lactam antibiotics intermediates. <i>Bioresource Technology</i> , 2010, 101, 331-336.	9.6	59
18	Physical location of the site for N-acetyl-L-glutamate, the allosteric activator of carbamoyl phosphate synthetase in the 20-kilodalton carboxy-terminal domain. <i>Biochemistry</i> , 1989, 28, 3070-3074.	2.5	54

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19	The Crystal Structure of Pyrococcus furiosus LUMP Kinase Provides Insight into Catalysis and Regulation in Microbial Pyrimidine Nucleotide Biosynthesis. <i>Journal of Molecular Biology</i> , 2005, 352, 438-454.	4.2	51
20	Mitochondrial Carbamoyl Phosphate Synthetase Activity in the Absence of N-Acetyl-l-glutamate. Mechanism of Activation by this Cofactor. <i>FEBS Journal</i> , 1983, 134, 337-343.	0.2	49
21	ATPase activity of biotin carboxylase provides evidence for initial activation of HCO ₃ ⁻ by ATP in the carboxylation of biotin. <i>Archives of Biochemistry and Biophysics</i> , 1986, 251, 465-470.	3.0	49
22	The 1.5 Å... resolution crystal structure of the carbamate kinase-like carbamoyl phosphate synthetase from the hyperthermophilic archaeon <i>Pyrococcus furiosus</i> , bound to ADP, confirms that this thermostable enzyme is a carbamate kinase, and provides insight into substrate binding and stability in carbamate kinases 1. Edited by R. Huber. <i>Journal of Molecular Biology</i> , 2000, 299, 463-476.	4.2	49
23	Characterization of genomic structure and polymorphisms in the human carbamyl phosphate synthetase I gene. <i>Gene</i> , 2003, 311, 51-57.	2.2	48
24	Arginine Biosynthesis in <i>Thermotoga maritima</i> : Characterization of the Arginine-Sensitive N-Acetyl-l-Glutamate Kinase. <i>Journal of Bacteriology</i> , 2004, 186, 6142-6149.	2.2	48
25	Carbamate kinase: New structural machinery for making carbamoyl phosphate, the common precursor of pyrimidines and arginine. <i>Protein Science</i> , 1999, 8, 934-940.	7.6	46
26	Mechanism of Carbamoyl-Phosphate Synthetase. Properties of the Two Binding Sites for ATP. <i>FEBS Journal</i> , 1979, 102, 521-530.	0.2	45
27	Understanding pyrroline-5-carboxylate synthetase deficiency: clinical, molecular, functional, and expression studies, structure-based analysis, and novel therapy with arginine. <i>Journal of Inherited Metabolic Disease</i> , 2012, 35, 761-776.	3.6	44
28	Site-directed Mutagenesis of <i>Escherichia coli</i> Acetylglutamate Kinase and Aspartokinase III Probes the Catalytic and Substrate-binding Mechanisms of these Amino Acid Kinase Family Enzymes and Allows Three-dimensional Modelling of Aspartokinase. <i>Journal of Molecular Biology</i> , 2003, 334, 459-476.	4.2	43
29	The PII-NAGK-PipX-NtcA Regulatory Axis of Cyanobacteria: A Tale of Changing Partners, Allosteric Effectors and Non-covalent Interactions. <i>Frontiers in Molecular Biosciences</i> , 2018, 5, 91.	3.5	43
30	Carbamoyl phosphate synthetase I of human liver. Purification, some properties and immunological cross-reactivity with the rat liver enzyme. <i>Biochimica Et Biophysica Acta - Biomembranes</i> , 1981, 659, 150-160.	2.6	42
31	<i>ALDH18A1</i> gene mutations cause dominant spastic paraplegia SPG9: loss of function effect and plausibility of a dominant negative mechanism. <i>Brain</i> , 2016, 139, e3-e3.	7.6	42
32	Relative frequency of mutations causing ornithine transcarbamylase deficiency in 78 families. <i>Human Genetics</i> , 1996, 97, 274-276.	3.8	41
33	The Course of Phosphorus in the Reaction of N-Acetyl-l-glutamate Kinase, Determined from the Structures of Crystalline Complexes, Including a Complex with an AlF ₄ ⁻ Transition State Mimic. <i>Journal of Molecular Biology</i> , 2003, 331, 231-244.	4.2	40
34	Estimation of the total number of disease-causing mutations in ornithine transcarbamylase (OTC) deficiency. Value of the OTC structure in predicting a mutation pathogenic potential. <i>Journal of Inherited Metabolic Disease</i> , 2007, 30, 217-226.	3.6	40
35	Mechanism of Carbamoyl-Phosphate Synthetase. Binding of ATP by the Rat-Liver Mitochondrial Enzyme. <i>FEBS Journal</i> , 1979, 93, 245-256.	0.2	39
36	Identification of seven novel missense mutations, two splice-site mutations, two microdeletions and a polymorphic amino acid substitution in the gene for ornithine transcarbamylase (OTC) in patients with OTC deficiency. <i>Human Mutation</i> , 2002, 19, 185-186.	2.5	39

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37	Understanding carbamoyl-phosphate synthetase I (CPS1) deficiency by using expression studies and structure-based analysis. <i>Human Mutation</i> , 2010, 31, 801-808.	2.5	35
38	SPR analysis of promoter binding of <i>Synechocystis</i> PCC6803 transcription factors NtcA and CRP suggests cross-talk and sheds light on regulation by effector molecules. <i>FEBS Letters</i> , 2014, 588, 2270-2276.	2.8	35
39	Binding of N-acetyl-L-glutamate to rat liver carbamoyl phosphate synthetase (ammonia). <i>FEBS Journal</i> , 1983, 135, 331-337.	0.2	34
40	Structure-function studies in carbamoyl phosphate synthetases. <i>Biochemical Society Transactions</i> , 1993, 21, 198-202.	3.4	34
41	Molecular Characterization of Carbamoyl-Phosphate Synthetase (CPS1) Deficiency Using Human Recombinant CPS1 as a Key Tool. <i>Human Mutation</i> , 2013, 34, 1149-1159.	2.5	34
42	Understanding Carbamoyl Phosphate Synthetase Deficiency: Impact of Clinical Mutations on Enzyme Functionality. <i>Journal of Molecular Biology</i> , 2005, 349, 127-141.	4.2	33
43	Mapping active site residues in glutamate-5-kinase. The substrate glutamate and the feed-back inhibitor proline bind at overlapping sites. <i>FEBS Letters</i> , 2006, 580, 6247-6253.	2.8	33
44	Photoaffinity Labeling with UMP of Lysine 992 of Carbamyl Phosphate Synthetase from <i>Escherichia coli</i> Allows Identification of the Binding Site for the Pyrimidine Inhibitor. <i>Biochemistry</i> , 1996, 35, 7247-7255.	2.5	32
45	Mechanism of activation of bicarbonate ion by mitochondrial carbamoyl-phosphate synthetase: formation of enzyme-bound adenosine diphosphate from the adenosine triphosphate that yields inorganic phosphate. <i>Biochemistry</i> , 1981, 20, 1969-1974.	2.5	30
46	Understanding carbamoyl phosphate synthetase (CPS1) deficiency by using the recombinantly purified human enzyme: Effects of CPS1 mutations that concentrate in a central domain of unknown function. <i>Molecular Genetics and Metabolism</i> , 2014, 112, 123-132.	1.1	30
47	Carbamate kinase from <i>Enterococcus faecalis</i> and <i>Enterococcus faecium</i> . Cloning of the genes, studies on the enzyme expressed in <i>Escherichia coli</i> , and sequence similarity with N-acetyl-L-glutamate kinase. <i>FEBS Journal</i> , 1998, 253, 280-291.	0.2	29
48	The Carbamoyl-phosphate Synthetase of <i>Pyrococcus furiosus</i> Is Enzymologically and Structurally a Carbamate Kinase. <i>Journal of Biological Chemistry</i> , 1999, 274, 16295-16303.	3.4	29
49	Prominent role of lysosomes in the proteolysis of rat liver mitochondria at neutral pH. <i>FEBS Letters</i> , 1977, 75, 281-284.	2.8	28
50	The Study of Carbamoyl Phosphate Synthetase 1 Deficiency Sheds Light on the Mechanism for Switching On/Off the Urea Cycle. <i>Journal of Genetics and Genomics</i> , 2015, 42, 249-260.	3.9	28
51	Dissection of <i>Escherichia coli</i> glutamate 5-kinase: Functional impact of the deletion of the PUA domain. <i>FEBS Letters</i> , 2005, 579, 6903-6908.	2.8	27
52	Studies on cyanobacterial protein PipY shed light on structure, potential functions, and vitamin B ₆ -dependent epilepsy. <i>FEBS Letters</i> , 2017, 591, 3431-3442.	2.8	27
53	Determination of N-acetyl-L-glutamate using high-performance liquid chromatography. <i>Analytical Biochemistry</i> , 1985, 146, 252-259.	2.4	26
54	Basis of Arginine Sensitivity of Microbial N -Acetyl- I -Glutamate Kinases: Mutagenesis and Protein Engineering Study with the <i>Pseudomonas aeruginosa</i> and <i>Escherichia coli</i> Enzymes. <i>Journal of Bacteriology</i> , 2008, 190, 3018-3025.	2.2	26

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55	Understanding N-Acetyl-L-Glutamate Synthase Deficiency: Mutational Spectrum, Impact of Clinical Mutations on Enzyme Functionality, and Structural Considerations. <i>Human Mutation</i> , 2016, 37, 679-694.	2.5	26
56	PipY, a Member of the Conserved COG0325 Family of PLP-Binding Proteins, Expands the Cyanobacterial Nitrogen Regulatory Network. <i>Frontiers in Microbiology</i> , 2017, 8, 1244.	3.5	26
57	A new case of arginase deficiency in a Spanish male. <i>Journal of Inherited Metabolic Disease</i> , 1986, 9, 393-397.	3.6	25
58	Structural insight on the control of urea synthesis: identification of the binding site for N-acetyl-L-glutamate, the essential allosteric activator of mitochondrial carbamoyl phosphate synthetase. <i>Biochemical Journal</i> , 2009, 424, 211-220.	3.7	25
59	Molecular mechanisms underlying large genomic deletions in ornithine transcarbamylase (<i>OTC</i>) gene. <i>Clinical Genetics</i> , 2009, 75, 457-464.	2.0	24
60	N-acetyl-L-glutamate in brain: Assay, levels, and regional and subcellular distribution. <i>Neurochemical Research</i> , 1991, 16, 787-794.	3.3	22
61	Site-directed mutagenesis of the regulatory domain of <i>Escherichia coli</i> carbamoyl phosphate synthetase identifies crucial residues for allosteric regulation and for transduction of the regulatory signals 1 Edited by A. R. Fersht. <i>Journal of Molecular Biology</i> , 2000, 299, 979-991.	4.2	22
62	The crystal structure of the cephalosporin deacetylating enzyme acetyl xylan esterase bound to paraoxon explains the low sensitivity of this serine hydrolase to organophosphate inactivation. <i>Biochemical Journal</i> , 2011, 436, 321-330.	3.7	22
63	Expanding the Cyanobacterial Nitrogen Regulatory Network: The GntR-Like Regulator PlmA Interacts with the PII-PipX Complex. <i>Frontiers in Microbiology</i> , 2016, 7, 1677.	3.5	22
64	Human Carbamoylphosphate Synthetase I. <i>Enzyme</i> , 1981, 26, 233-239.	0.7	21
65	Insight into vitamin B ₆ -dependent epilepsy due to <i>PLPBP</i> (previously <i>PROSC</i>) missense mutations. <i>Human Mutation</i> , 2018, 39, 1002-1013.	2.5	21
66	Mechanism of arginine regulation of acetylglutamate synthase, the first enzyme of arginine synthesis. <i>FEBS Letters</i> , 2009, 583, 202-206.	2.8	20
67	" ¹ H-Pyrroline-5-carboxylate synthetase deficiency: An emergent multifaceted urea cycle-related disorder. <i>Journal of Inherited Metabolic Disease</i> , 2020, 43, 657-670.	3.6	20
68	Treating urea cycle defects. <i>Nature</i> , 1981, 292, 496-496.	27.8	19
69	Substrate Binding and Catalysis in Carbamate Kinase Ascertained by Crystallographic and Site-Directed Mutagenesis Studies: Movements and Significance of a Unique Globular Subdomain of This Key Enzyme for Fermentative ATP Production in Bacteria. <i>Journal of Molecular Biology</i> , 2010, 397, 1261-1275.	4.2	19
70	Photoaffinity Labeling with the Activator IMP and Site-Directed Mutagenesis of Histidine 995 of Carbamoyl Phosphate Synthetase from <i>Escherichia coli</i> Demonstrate That the Binding Site for IMP Overlaps with That for the Inhibitor UMP. <i>Biochemistry</i> , 1999, 38, 3910-3917.	2.5	18
71	Insight on an Arginine Synthesis Metabolon from the Tetrameric Structure of Yeast Acetylglutamate Kinase. <i>PLoS ONE</i> , 2012, 7, e34734.	2.5	18
72	Affinity Cleavage of Carbamoyl-Phosphate Synthetase I Localizes Regions of the Enzyme Interacting with the Molecule of ATP that Phosphorylates Carbamate. <i>FEBS Journal</i> , 1995, 229, 377-384.	0.2	18

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73	Location of the Binding Site for the Allosteric Activator IMP in the COOH-Terminal Domain of Escherichia coli Carbamyl Phosphate Synthetase. Biochemical and Biophysical Research Communications, 1994, 203, 1083-1089.	2.1	17
74	Localization of the site for the nucleotide effectors of Escherichia coli carbamoyl phosphate synthetase using site-directed mutagenesis. FEBS Letters, 1999, 446, 133-136.	2.8	16
75	Carbamate kinase can replace in vivo carbamoyl phosphate synthetase. Implications for the evolution of carbamoyl phosphate biosynthesis. FEBS Letters, 2000, 484, 261-264.	2.8	16
76	Site-directed mutagenesis studies of acetylglutamate synthase delineate the site for the arginine inhibitor. FEBS Letters, 2008, 582, 1081-1086.	2.8	16
77	Ligand binding specificity of RutR, a member of the TetR family of transcription regulators in Escherichia coli. FEBS Open Bio, 2015, 5, 76-84.	2.3	16
78	Synthesis of carbamoyl phosphate by carbamoyl phosphate synthetase I in the absence of acetylglutamate. Activation of the enzyme by cryoprotectants. Biochemical and Biophysical Research Communications, 1981, 99, 1131-1137.	2.1	15
79	Orotic Aciduria Due to Arginine Deprivation: Changes in the Levels of Carbamoyl Phosphate and of Other Urea Cycle Intermediates in Mouse Liver. Journal of Nutrition, 1989, 119, 1188-1195.	2.9	15
80	A structure-reactivity study of the binding of acetylglutamate to carbamoyl phosphate synthetase I. FEBS Journal, 1990, 188, 47-53.	0.2	15
81	Demonstration of the spf-ash mutation in Spanish patients with ornithine transcarbamylase deficiency of moderate severity. Human Genetics, 1995, 95, 183-6.	3.8	15
82	Two Crystal Structures of Escherichia coli N-Acetyl-L-Glutamate Kinase Demonstrate the Cycling between Open and Closed Conformations. Journal of Molecular Biology, 2010, 399, 476-490.	4.2	15
83	Structure of AmtR, the global nitrogen regulator of Corynebacterium glutamicum, in free and DNA-bound forms. FEBS Journal, 2016, 283, 1039-1059.	4.7	15
84	Enzymatic HCO ₃ ²⁻ fixation: A common mechanism for all enzymes involved?. Bioscience Reports, 1986, 6, 335-347.	2.4	14
85	Human growth plate development in the fetal and neonatal period. Journal of Orthopaedic Research, 1992, 10, 62-71.	2.3	14
86	Mechanism of carbamoyl phosphate synthetase from Escherichia coli. Binding of the ATP molecules used in the reaction and sequestration by the enzyme of the ATP molecule that yields carbamoyl phosphate. FEBS Journal, 1998, 255, 262-270.	0.2	14
87	Glutamate-5-kinase from Escherichia coli: gene cloning, overexpression, purification and crystallization of the recombinant enzyme and preliminary X-ray studies. Acta Crystallographica Section D: Biological Crystallography, 2004, 60, 2091-2094.	2.5	14
88	Citrin deficiency in a Romanian child living in Spain highlights the worldwide distribution of this defect and illustrates the value of nutritional therapy. Molecular Genetics and Metabolism, 2013, 110, 181-183.	1.1	14
89	P5CS expression study in a new family with ALDH18A1-associated hereditary spastic paraplegia SPG9. Annals of Clinical and Translational Neurology, 2019, 6, 1533-1540.	3.7	14
90	Functional and structural characterization of PII-like protein CutA does not support involvement in heavy metal tolerance and hints at a small molecule carrying/signaling role. FEBS Journal, 2021, 288, 1142-1162.	4.7	14

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91	Activation of carbamoyl phosphate synthetase by cryoprotectants. <i>Molecular and Cellular Biochemistry</i> , 1983, 53-54, 279-98.	3.1	13
92	The structure of a PII signaling protein from a halophilic archaeon reveals novel traits and high salt adaptations. <i>FEBS Journal</i> , 2014, 281, 3299-3314.	4.7	13
93	Disorders of the Urea Cycle and Related Enzymes. , 2016, , 295-308.		13
94	Limited proteolysis reveals low-affinity binding of N-acetyl-L-glutamate to rat-liver carbamoyl-phosphate synthetase (ammonia). <i>FEBS Journal</i> , 1987, 165, 163-169.	0.2	12
95	Structures of collagen IV globular domains: insight into associated pathologies, folding and network assembly. <i>IUCr</i> , 2018, 5, 765-779.	2.2	12
96	Identification of a cytogenetic deletion and of four novel mutations (Q69X, I172F, G188V, G197R) affecting the gene for ornithine transcarbamylase (OTC) in Spanish patients with OTC deficiency. <i>Human Mutation</i> , 1999, 14, 352-353.	2.5	11
97	Mechanism of oligomerization of <i>Escherichia coli</i> carbamoyl phosphate synthetase and modulation by the allosteric effectors. A site-directed mutagenesis study. <i>FEBS Letters</i> , 2002, 511, 6-10.	2.8	11
98	Hyperammonemias and Related Disorders. , 2014, , 47-62.		11
99	N-Acetyl-L-glutamate kinase from <i>Escherichia coli</i> : cloning of the gene, purification and crystallization of the recombinant enzyme and preliminary X-ray analysis of the free and ligand-bound forms. <i>Acta Crystallographica Section D: Biological Crystallography</i> , 1999, 55, 1350-1352.	2.5	10
100	Carbamoyl-phosphate synthetase I. Kinetics of binding and dissociation of acetylglutamate and of activation and deactivation. <i>FEBS Journal</i> , 1988, 171, 615-622.	0.2	9
101	Carbamoyl Phosphate Synthetase, Ornithine Transcarbamylase, and Aspartate Transcarbamylase Activities in the Pea Ovary. <i>Plant Physiology</i> , 1989, 90, 1565-1569.	4.8	9
102	A splicing mutation, a nonsense mutation (Y167X) and two missense mutations (I159T and A209V) in Spanish patients with ornithine transcarbamylase deficiency. <i>Human Genetics</i> , 1995, 96, 549-51.	3.8	9
103	Molecular Physiology of Phosphoryl Group Transfer from Carbamoyl Phosphate by a Hyperthermophilic Enzyme at Low Temperature. <i>Biochemistry</i> , 2002, 41, 3916-3924.	2.5	9
104	First-time crystallization and preliminary X-ray crystallographic analysis of a bacterial-archaeal type UMP kinase, a key enzyme in microbial pyrimidine biosynthesis. <i>Biochimica Et Biophysica Acta - Proteins and Proteomics</i> , 2005, 1747, 271-275.	2.3	9
105	Functional Dissection of N -Acetylglutamate Synthase (ArgA) of <i>Pseudomonas aeruginosa</i> and Restoration of Its Ancestral N -Acetylglutamate Kinase Activity. <i>Journal of Bacteriology</i> , 2012, 194, 2791-2801.	2.2	9
106	Inactivation of carbamoyl phosphate synthetase (ammonia) by elastase as a probe to investigate binding of the substrates. <i>Biochemical and Biophysical Research Communications</i> , 1983, 117, 238-244.	2.1	8
107	[21] Carbamoyl phosphate synthesis: Carbamate kinase from <i>Pyrococcus furiosus</i> . <i>Methods in Enzymology</i> , 2001, 331, 236-247.	1.0	8
108	Recurrence of carbamoyl phosphate synthetase 1 (CPS1) deficiency in Turkish patients: Characterization of a founder mutation by use of recombinant CPS1 from insect cells expression. <i>Molecular Genetics and Metabolism</i> , 2014, 113, 267-273.	1.1	8

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109	The PipX Protein, When Not Bound to Its Targets, Has Its Signaling C-Terminal Helix in a Flexed Conformation. <i>Biochemistry</i> , 2017, 56, 3211-3224.	2.5	8
110	Autoradiographic evidence of increased incorporation of aspartate and of carbamoyl aspartate in fibroblasts from a Lesch-Nyhan patient. <i>Biochemical and Biophysical Research Communications</i> , 1979, 90, 333-337.	2.1	7
111	High-performance liquid chromatographic assay of argininosuccinate: its application in argininosuccinic aciduria and in normal man. <i>Journal of Inherited Metabolic Disease</i> , 1986, 9, 31-38.	3.6	7
112	Inactivation of mitochondrial carbamoyl phosphate synthetase induced by ascorbate, oxygen, and Fe ³⁺ in the presence of acetylglutamate: Protection by ATP and HCO ³⁻ and lack of inactivation of ornithine transcarbamylase. <i>Archives of Biochemistry and Biophysics</i> , 1987, 258, 342-350.	3.0	7
113	Crystallization, Characterization and Preliminary Crystallographic Studies of Carbamate Kinase of <i>Streptococcus faecium</i> . <i>Journal of Molecular Biology</i> , 1994, 235, 1345-1347.	4.2	7
114	Missense mutations in codon 225 of ornithine transcarbamylase (OTC) result in decreased amounts of OTC protein: A hypothesis on the molecular mechanism of the OTC deficiency. <i>Journal of Inherited Metabolic Disease</i> , 1997, 20, 769-777.	3.6	7
115	Influence of dose and age on the response of the allopurinol test for ornithine carbamoyltransferase deficiency in control infants. <i>Journal of Inherited Metabolic Disease</i> , 2000, 23, 662-668.	3.6	7
116	Towards structural understanding of feedback control of arginine biosynthesis: cloning and expression of the gene for the arginine-inhibited N-acetyl-L-glutamate kinase from <i>Pseudomonas aeruginosa</i> , purification and crystallization of the recombinant enzyme and preliminary X-ray studies. <i>Acta Crystallographica Section D: Biological Crystallography</i> , 2002, 58, 1045-1047.	2.5	7
117	Carbamoylglutamate-responsive carbamoyl phosphate synthetase 1 (CPS1) deficiency: A patient with a novel CPS1 mutation and an experimental study on the mutation's effects. <i>JMD Reports</i> , 2019, 48, 36-44.	1.5	7
118	Discovery of 3H-pyrrolo[2,3-c]quinolines with activity against <i>Mycobacterium tuberculosis</i> by allosteric inhibition of the glutamate-5-kinase enzyme. <i>European Journal of Medicinal Chemistry</i> , 2022, 232, 114206.	5.5	7
119	Fibronectin in Bronchoalveolar Lavage Fluid in Lung Cancer: Tumor or Inflammatory Marker?. <i>Respiration</i> , 1998, 65, 178-182.	2.6	6
120	H intragenic polymorphisms and haplotype analysis in the ornithine transcarbamylase (OTC) gene and their relevance for tracking the inheritance of OTC deficiency. <i>Human Mutation</i> , 2002, 20, 407-408.	2.5	6
121	Effects of T-loop modification on the PII signalling protein: structure of uridylylated <i>Escherichia coli</i> GlnB bound to ATP. <i>Environmental Microbiology Reports</i> , 2017, 9, 290-299.	2.4	6
122	Insight on molecular pathogenesis and pharmacochaperoning potential in phosphomannomutase 2 deficiency, provided by novel human phosphomannomutase 2 structures. <i>Journal of Inherited Metabolic Disease</i> , 2022, 45, 318-333.	3.6	6
123	Activation of carbamoyl phosphate synthetase from <i>Escherichia coli</i> by glycerol. <i>Biochemical and Biophysical Research Communications</i> , 1982, 107, 1400-1405.	2.1	5
124	Neonatal citrullinaemia with satisfactory mental development. <i>European Journal of Pediatrics</i> , 1991, 150, 730-731.	2.7	5
125	Mechanism of allosteric modulation of <i>Escherichia coli</i> carbamoyl phosphate synthetase probed by site-directed mutagenesis of ornithine site residues. <i>FEBS Letters</i> , 2002, 514, 323-328.	2.8	5
126	Mutant alleles associated with late-onset ornithine transcarbamylase deficiency in male patients have recurrently arisen and have been retained in some populations. <i>Journal of Human Genetics</i> , 2010, 55, 18-22.	2.3	5

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127	Solvatochromic and pH-Sensitive Fluorescent Membrane Probes for Imaging of Live Cells. ACS Chemical Neuroscience, 2021, 12, 719-734.	3.5	5
128	New Insight into the Transcarbamylase Family: The Structure of Putrescine Transcarbamylase, a Key Catalyst for Fermentative Utilization of Agmatine. PLoS ONE, 2012, 7, e31528.	2.5	5
129	Influence of anions on the activation of carbamoyl phosphate synthetase (ammonia) by acetylglutamate: Implications for the activation of the enzyme in the mitochondria. Archives of Biochemistry and Biophysics, 1991, 288, 414-420.	3.0	4
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