

Hiroki Morizono

List of Publications by Year in descending order

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71
papers

3,349
citations

147801

31
h-index

155660

55
g-index

76
all docs

76
docs citations

76
times ranked

3924
citing authors

#	ARTICLE	IF	CITATIONS
1	A dual AAV system enables the Cas9-mediated correction of a metabolic liver disease in newborn mice. <i>Nature Biotechnology</i> , 2016, 34, 334-338.	17.5	476
2	Adeno-Associated Virus Antibody Profiles in Newborns, Children, and Adolescents. <i>Vaccine Journal</i> , 2011, 18, 1586-1588.	3.1	269
3	Mutations and polymorphisms in the human ornithine transcarbamylase (<i>OTC</i>) gene. <i>Human Mutation</i> , 2006, 27, 626-632.	2.5	184
4	Clinical Characterization and Prediction of Clinical Severity of SARS-CoV-2 Infection Among US Adults Using Data From the US National COVID Cohort Collaborative. <i>JAMA Network Open</i> , 2021, 4, e2116901.	5.9	179
5	Baseline human gut microbiota profile in healthy people and standard reporting template. <i>PLoS ONE</i> , 2019, 14, e0206484.	2.5	133
6	The Pleiotropic Effects of Natural AAV Infections on Liver-directed Gene Transfer in Macaques. <i>Molecular Therapy</i> , 2010, 18, 126-134.	8.2	123
7	Mutations and polymorphisms in the human ornithine transcarbamylase gene. <i>Human Mutation</i> , 2002, 19, 93-107.	2.5	120
8	Genotypeâ€“Phenotype Correlations in Ornithine Transcarbamylase Deficiency: A Mutation Update. <i>Journal of Genetics and Genomics</i> , 2015, 42, 181-194.	3.9	111
9	Systematic Evaluation of AAV Vectors for Liver directed Gene Transfer in Murine Models. <i>Molecular Therapy</i> , 2010, 18, 118-125.	8.2	110
10	Long-Term Correction of Ammonia Metabolism and Prolonged Survival in Ornithine Transcarbamylase-Deficient Mice Following Liver-Directed Treatment with Adeno-associated Viral Vectors. <i>Molecular Therapy</i> , 2006, 14, 25-33.	8.2	76
11	Cloning and expression of the human N-acetylglutamate synthase gene. <i>Biochemical and Biophysical Research Communications</i> , 2002, 299, 581-586.	2.1	74
12	1.85-Å... Resolution Crystal Structure of Human Ornithine Transcarbamoylase Complexed with N-Phosphonacetyl-l-ornithine. <i>Journal of Biological Chemistry</i> , 1998, 273, 34247-34254.	3.4	73
13	Restoration of ureagenesis in N-acetylglutamate synthase deficiency by N-carbamylglutamate. <i>Journal of Pediatrics</i> , 2004, 145, 552-554.	1.8	58
14	Identification, cloning and expression of the mouse N-acetylglutamate synthase gene. <i>Biochemical Journal</i> , 2002, 364, 825-831.	3.7	52
15	Null mutations in the N-acetylglutamate synthase gene associated with acute neonatal disease and hyperammonemia. <i>Human Genetics</i> , 2003, 112, 364-368.	3.8	49
16	N-acetylglutamate synthase: structure, function and defects. <i>Molecular Genetics and Metabolism</i> , 2010, 100, S13-S19.	1.1	49
17	Human ornithine transcarbamylase: crystallographic insights into substrate recognition and conformational changes. <i>Biochemical Journal</i> , 2001, 354, 501-509.	3.7	48
18	Genetic and Clinical Heterogeneity in eIF2B-Related Disorder. <i>Journal of Child Neurology</i> , 2008, 23, 205-215.	1.4	46

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19	Preclinical evaluation of a clinical candidate AAV8 vector for ornithine transcarbamylase (OTC) deficiency reveals functional enzyme from each persisting vector genome. <i>Molecular Genetics and Metabolism</i> , 2012, 105, 203-211.	1.1	46
20	A mutation-independent CRISPR-Cas9-mediated gene targeting approach to treat a murine model of ornithine transcarbamylase deficiency. <i>Science Advances</i> , 2020, 6, eaax5701.	10.3	44
21	Acetylornithine Transcarbamylase: a Novel Enzyme in Arginine Biosynthesis. <i>Journal of Bacteriology</i> , 2006, 188, 2974-2982.	2.2	42
22	Expression, purification, and characterization of recombinant human glutamine synthetase. <i>Biochemical Journal</i> , 1997, 328, 159-163.	3.7	41
23	Mammalian N-acetylglutamate synthase. <i>Molecular Genetics and Metabolism</i> , 2004, 81, 4-11.	1.1	39
24	Crystal Structure of N-Acetylornithine Transcarbamylase from <i>Xanthomonas campestris</i> . <i>Journal of Biological Chemistry</i> , 2005, 280, 14366-14369.	3.4	39
25	AAV gene therapy corrects OTC deficiency and prevents liver fibrosis in aged OTC-knock out heterozygous mice. <i>Molecular Genetics and Metabolism</i> , 2017, 120, 299-305.	1.1	39
26	Sustained correction of OTC deficiency in spfash mice using optimized self-complementary AAV2/8 vectors. <i>Gene Therapy</i> , 2012, 19, 404-410.	4.5	38
27	Human ornithine transcarbamylase: crystallographic insights into substrate recognition and conformational changes. <i>Biochemical Journal</i> , 2001, 354, 501.	3.7	38
28	Expression, purification and kinetic characterization of wild-type human ornithine transcarbamylase and a recurrent mutant that produces "late onset" hyperammonaemia. <i>Biochemical Journal</i> , 1997, 322, 625-631.	3.7	37
29	Late onset N-acetylglutamate synthase deficiency caused by hypomorphic alleles. <i>Human Mutation</i> , 2005, 25, 293-298.	2.5	37
30	Disease-causing mutations in the promoter and enhancer of the ornithine transcarbamylase gene. <i>Human Mutation</i> , 2018, 39, 527-536.	2.5	37
31	Mutations and polymorphisms in the human N-acetylglutamate synthase (NAGS) gene. <i>Human Mutation</i> , 2007, 28, 754-759.	2.5	36
32	Identification of "private" mutations in patients with ornithine transcarbamylase deficiency. <i>Journal of Inherited Metabolic Disease</i> , 1997, 20, 525-527.	3.6	35
33	Crystal structure of human ornithine transcarbamylase complexed with carbamoyl phosphate and L-norvaline at 1.9 Å resolution. <i>Journal of Biological Chemistry</i> , 2000, 275, 271-277.		35
34	Biochemical properties of recombinant human and mouse N-acetylglutamate synthase. <i>Molecular Genetics and Metabolism</i> , 2006, 87, 226-232.	1.1	34
35	The Crystal Structure of N-Acetyl-L-glutamate Synthase from <i>Neisseria gonorrhoeae</i> Provides Insights into Mechanisms of Catalysis and Regulation. <i>Journal of Biological Chemistry</i> , 2008, 283, 7176-7184.	3.4	33
36	Design and Implementation of the Hepatorenal Fibrocystic Disease Core Center Clinical Database: A Centralized Resource for Characterizing Autosomal Recessive Polycystic Kidney Disease and Other Hepatorenal Fibrocystic Diseases. <i>Frontiers in Pediatrics</i> , 2017, 5, 80.	1.9	31

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37	Inversion of allosteric effect of arginine on N-acetylglutamate synthase, a molecular marker for evolution of tetrapods. <i>BMC Biochemistry</i> , 2008, 9, 24.	4.4	30
38	Enabling precision medicine via standard communication of HTS provenance, analysis, and results. <i>PLoS Biology</i> , 2018, 16, e3000099.	5.6	29
39	A novel bifunctional N-acetylglutamate synthase-kinase from <i>Xanthomonas campestris</i> that is closely related to mammalian N-acetylglutamate synthase. <i>BMC Biochemistry</i> , 2007, 8, 4.	4.4	28
40	Dysregulated calcium homeostasis prevents plasma membrane repair in Anoctamin 5/TMEM16E-deficient patient muscle cells. <i>Cell Death Discovery</i> , 2019, 5, 118.	4.7	28
41	Mechanism of Allosteric Inhibition of N-Acetyl-L-glutamate Synthase by L-Arginine. <i>Journal of Biological Chemistry</i> , 2009, 284, 4873-4880.	3.4	27
42	Canine Heparan Sulfate Sulfamidase and the Molecular Pathology Underlying Sanfilippo Syndrome Type A in Dachshunds. <i>Genomics</i> , 2000, 68, 80-84.	2.9	25
43	Crystal Structure of a Transcarbamylase-like Protein from the Anaerobic Bacterium <i>Bacteroides fragilis</i> at 2.0Å... Resolution. <i>Journal of Molecular Biology</i> , 2002, 320, 899-908.	4.2	23
44	Structure and Catalytic Mechanism of a Novel N-Succinyl-L-ornithine Transcarbamylase in Arginine Biosynthesis of <i>Bacteroides fragilis</i> . <i>Journal of Biological Chemistry</i> , 2006, 281, 20623-20631.	3.4	22
45	A novel biochemically salvageable animal model of hyperammonemia devoid of N-acetylglutamate synthase. <i>Molecular Genetics and Metabolism</i> , 2012, 106, 160-168.	1.1	21
46	Molecular Recognition by Ornithine and Aspartate Transcarbamylases. <i>Accounts of Chemical Research</i> , 1999, 32, 885-894.	15.6	19
47	A single mutation in the active site swaps the substrate specificity of N-acetyl-L-ornithine transcarbamylase and N-succinyl-L-ornithine transcarbamylase. <i>Protein Science</i> , 2007, 16, 1689-1699.	7.6	17
48	Criticality: A New Concept of Severity of Illness for Hospitalized Children. <i>Pediatric Critical Care Medicine</i> , 2021, 22, e33-e43.	0.5	17
49	â€˜Late Onsetâ€™ Ornithine Transcarbamylase Deficiency: Function of Three Purified Recombinant Mutant Enzymes. <i>Human Molecular Genetics</i> , 1997, 6, 963-968.	2.9	16
50	Structures of N -acetylornithine transcarbamoylase from <i>Xanthomonas campestris</i> complexed with substrates and substrate analogs imply mechanisms for substrate binding and catalysis. <i>Proteins: Structure, Function and Bioinformatics</i> , 2006, 64, 532-542.	2.6	15
51	Genome-wide profiling of differentially spliced mRNAs in human fetal cortical tissue exposed to alcohol. <i>Alcohol</i> , 2017, 62, 1-9.	1.7	15
52	Predicting the severity of motor neuron disease progression using electronic health record data with a cloud computing Big Data approach. , 2014, 2014, .		14
53	Expression, purification, crystallization and preliminary X-ray crystallographic studies of a novel acetylcitrulline deacetylase from <i>Xanthomonas campestris</i> . <i>Acta Crystallographica Section F: Structural Biology Communications</i> , 2005, 61, 676-679.	0.7	11
54	The use of yeast mitochondria to study the properties of wild-type and mutant human mitochondrial ornithine transporter. <i>Molecular Genetics and Metabolism</i> , 2005, 86, 431-440.	1.1	11

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55	Severity Trajectories of Pediatric Inpatients Using the Criticality Index. <i>Pediatric Critical Care Medicine</i> , 2021, 22, e19-e32.	0.5	11
56	Expression of Wild-Type and Mutant Human Ornithine Transcarbamylase Genes in Chinese Hamster Ovary Cells and Lack of Dominant Negative Effect of R141Q and R40H Mutants. <i>Pediatric Research</i> , 2000, 48, 842-846.	2.3	10
57	Expression, crystallization and preliminary crystallographic studies of a novel bifunctional N-acetylglutamate synthase/kinase from <i>Xanthomonas campestris</i> homologous to vertebrate N-acetylglutamate synthase. <i>Acta Crystallographica Section F: Structural Biology Communications</i> , 2006, 62, 1218-1222.	0.7	10
58	Mitochondrial Enzymes of the Urea Cycle Cluster at the Inner Mitochondrial Membrane. <i>Frontiers in Physiology</i> , 2020, 11, 542950.	2.8	10
59	Association of Congenital and Acquired Cardiovascular Conditions With COVID-19 Severity Among Pediatric Patients in the US. <i>JAMA Network Open</i> , 2022, 5, e2211967.	5.9	8
60	The clinically variable R40H mutant ornithine carbamoyltransferase shows cytosolic degradation of the precursor protein in CHO cells. <i>Journal of Inherited Metabolic Disease</i> , 2001, 24, 614-622.	3.6	7
61	Gene delivery corrects N-acetylglutamate synthase deficiency and enables insights in the physiological impact of L-arginine activation of N-acetylglutamate synthase. <i>Scientific Reports</i> , 2021, 11, 3580.	3.3	6
62	Crystallization and preliminary X-ray crystallographic studies of wild-type human ornithine transcarbamylase and two naturally occurring mutants at position 277. <i>Acta Crystallographica Section D: Biological Crystallography</i> , 2001, 57, 719-721.	2.5	2
63	Standardized Health data and Research Exchange (SHaRE): promoting a learning health system. <i>JAMIA Open</i> , 2022, 5, ooab120.	2.0	2
64	Validation of a computational phenotype for finding patients eligible for genetic testing for pathogenic PTEN variants across three centers. <i>Journal of Neurodevelopmental Disorders</i> , 2022, 14, 24.	3.1	2
65	Facilitating orphan drug development: Proceedings of the TREAT-NMD International Conference, December 2015, Washington, DC, USA. <i>Neuromuscular Disorders</i> , 2017, 27, 693-701.	0.6	1
66	Predicting Future Care Requirements Using Machine Learning for Pediatric Intensive and Routine Care Inpatients. , 2021, 3, e0505.		1
67	174. Liver Fibrosis in Aged OTC-KO Heterozygotes and Successful Correction by AAV8-Mediated Gene Therapy. <i>Molecular Therapy</i> , 2015, 23, S69.	8.2	0
68	481. CRISPR/Cas9-Mediated In Vivo Genome Editing to Correct the OTC spfash Mutation in Newborn Mice. <i>Molecular Therapy</i> , 2016, 24, S190-S191.	8.2	0
69	Characterization of novel microbial transcarbamylases. <i>FASEB Journal</i> , 2007, 21, A1013.	0.5	0
70	Natural Variation in Expression of Urea Cycle Genes. <i>FASEB Journal</i> , 2007, 21, A664.	0.5	0
71	The Interaction of N-acetylglutamate Synthase with Other Mitochondrial Urea Cycle Enzymes. <i>FASEB Journal</i> , 2010, 24, 658.4.	0.5	0