Hiroki Morizono

List of Publications by Year
in descending order

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71	3,349	147801	155660
papers	citations	31 h-index	55 g-index

76	76	76	3924
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	Standardized Health data and Research Exchange (SHaRE): promoting a learning health system. JAMIA Open, 2022, 5, ooab120.	2.0	2
2	Validation of a computational phenotype for finding patients eligible for genetic testing for pathogenic PTEN variants across three centers. Journal of Neurodevelopmental Disorders, 2022, 14, 24.	3.1	2
3	Association of Congenital and Acquired Cardiovascular Conditions With COVID-19 Severity Among Pediatric Patients in the US. JAMA Network Open, 2022, 5, e2211967.	5.9	8
4	Gene delivery corrects N-acetylglutamate synthase deficiency and enables insights in the physiological impact of L-arginine activation of N-acetylglutamate synthase. Scientific Reports, 2021, 11, 3580.	3.3	6
5	Clinical Characterization and Prediction of Clinical Severity of SARS-CoV-2 Infection Among US Adults Using Data From the US National COVID Cohort Collaborative. JAMA Network Open, 2021, 4, e2116901.	5.9	179
6	Criticality: A New Concept of Severity of Illness for Hospitalized Children. Pediatric Critical Care Medicine, 2021, 22, e33-e43.	0.5	17
7	Severity Trajectories of Pediatric Inpatients Using the Criticality Index. Pediatric Critical Care Medicine, 2021, 22, e19-e32.	0.5	11
8	Predicting Future Care Requirements Using Machine Learning for Pediatric Intensive and Routine Care Inpatients. , 2021, 3, e0505.		1
9	A mutation-independent CRISPR-Cas9â€“mediated gene targeting approach to treat a murine model of ornithine transcarbamylase deficiency. Science Advances, 2020, 6, eaax5701.	10.3	44
10	Mitochondrial Enzymes of the Urea Cycle Cluster at the Inner Mitochondrial Membrane. Frontiers in Physiology, 2020, 11, 542950.	2.8	10
11	Dysregulated calcium homeostasis prevents plasma membrane repair in Anoctamin 5/TMEM16E-deficient patient muscle cells. Cell Death Discovery, 2019, 5, 118.	4.7	28
12	Baseline human gut microbiota profile in healthy people and standard reporting template. PLoS ONE, 2019, 14, e0206484.	2.5	133
13	Disease-causing mutations in the promoter and enhancer of the ornithine transcarbamylase gene. Human Mutation, 2018, 39, 527-536.	2.5	37
14	Enabling precision medicine via standard communication of HTS provenance, analysis, and results. PLoS Biology, 2018, 16, e3000099.	5.6	29
15	AAV gene therapy corrects OTC deficiency and prevents liver fibrosis in aged OTC-knock out heterozygous mice. Molecular Genetics and Metabolism, 2017, 120, 299-305.	1.1	39
16	Genome-wide profiling of differentially spliced mRNAs in human fetal cortical tissue exposed to alcohol. Alcohol, 2017, 62, 1-9.	1.7	15
17	Facilitating orphan drug development: Proceedings of the TREAT-NMD International Conference, December 2015, Washington, DC, USA. Neuromuscular Disorders, 2017, 27, 693-701.	0.6	1
18	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. Frontiers in Pediatrics, 2017, 5, 80.	1.9	31

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19	481. CRISPR/Cas9-Mediated In Vivo Genome Editing to Correct the OTC spfash Mutation in Newborn Mice. Molecular Therapy, 2016, 24, S190-S191.	8.2	0
20	A dual AAV system enables the Cas9-mediated correction of a metabolic liver disease in newborn mice. Nature Biotechnology, 2016, 34, 334-338.	17.5	476
21	174. Liver Fibrosis in Aged OTC-KO Heterozygotes and Successful Correction by AAV8-Mediated Gene Therapy. Molecular Therapy, 2015, 23, S69.	8.2	0
22	Genotypeâ€“Phenotype Correlations in Ornithine Transcarbamylase Deficiency: A Mutation Update. Journal of Genetics and Genomics, 2015, 42, 181-194.	3.9	111
23	Predicting the severity of motor neuron disease progression using electronic health record data with a cloud computing Big Data approach. , 2014, 2014, .		14
24	Sustained correction of OTC deficiency in spfash mice using optimized self-complementary AAV2/8 vectors. Gene Therapy, 2012, 19, 404-410.	4.5	38
25	Preclinical evaluation of a clinical candidate AAV8 vector for ornithine transcarbamylase (OTC) deficiency reveals functional enzyme from each persisting vector genome. Molecular Genetics and Metabolism, 2012, 105, 203-211.	1.1	46
26	A novel biochemically salvageable animal model of hyperammonemia devoid of N-acetylglutamate synthase. Molecular Genetics and Metabolism, 2012, 106, 160-168.	1.1	21
27	Adeno-Associated Virus Antibody Profiles in Newborns, Children, and Adolescents. Vaccine Journal, 2011, 18, 1586-1588.	3.1	269
28	Systematic Evaluation of AAV Vectors for Liver directed Gene Transfer in Murine Models. Molecular Therapy, 2010, 18, 118-125.	8.2	110
29	The Pleiotropic Effects of Natural AAV Infections on Liver-directed Gene Transfer in Macaques. Molecular Therapy, 2010, 18, 126-134.	8.2	123
30	N-acetylglutamate synthase: structure, function and defects. Molecular Genetics and Metabolism, 2010, 100, S13-S19.	1.1	49
31	The Interaction of Nâ€Acetylglutamate Synthase with Other Mitochondrial Urea Cycle Enzymes. FASEB Journal, 2010, 24, 658.4.	0.5	0
32	Mechanism of Allosteric Inhibition of N-Acetyl-L-glutamate Synthase by L-Arginine. Journal of Biological Chemistry, 2009, 284, 4873-4880.	3.4	27
33	Inversion of allosteric effect of arginine on N-acetylglutamate synthase, a molecular marker for evolution of tetrapods. BMC Biochemistry, 2008, 9, 24.	4.4	30
34	Genetic and Clinical Heterogeneity in eIF2B-Related Disorder. Journal of Child Neurology, 2008, 23, 205-215.	1.4	46
35	The Crystal Structure of N-Acetyl-L-glutamate Synthase from Neisseria gonorrhoeae Provides Insights into Mechanisms of Catalysis and Regulation. Journal of Biological Chemistry, 2008, 283, 7176-7184.	3.4	33
36	Mutations and polymorphisms in the human N-acetylglutamate synthase (NAGS) gene. Human Mutation, 2007, 28, 754-759.	2.5	36

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37	A novel bifunctional N-acetylglutamate synthase-kinase from Xanthomonas campestris that is closely related to mammalian N-acetylglutamate synthase. BMC Biochemistry, 2007, 8, 4.	4.4	28
38	A single mutation in the active site swaps the substrate specificity ofN-acetyl-L-ornithine transcarbamylase andN-succinyl-L-ornithine transcarbamylase. Protein Science, 2007, 16, 1689-1699.	7.6	17
39	Characterization of novel microbial transcarbamylases. FASEB Journal, 2007, 21, A1013.	0.5	0
40	Natural Variation in Expression of Urea Cycle Genes. FASEB Journal, 2007, 21, A664.	0.5	0
41	Biochemical properties of recombinant human and mouse N-acetylglutamate synthase. Molecular Genetics and Metabolism, 2006, 87, 226-232.	1.1	34
42	Expression, crystallization and preliminary crystallographic studies of a novel bifunctionalN-acetylglutamate synthase/kinase fromXanthomonas campestrishomologous to vertebrateN-acetylglutamate synthase. Acta Crystallographica Section F: Structural Biology Communications, 2006, 62, 1218-1222.	0.7	10
43	Structures of N -acetylornithine transcarbamoylase from Xanthomonas campestris complexed with substrates and substrate analogs imply mechanisms for substrate binding and catalysis. Proteins: Structure, Function and Bioinformatics, 2006, 64, 532-542.	2.6	15
44	Mutations and polymorphisms in the human ornithine transcarbamylase (OTC) gene. Human Mutation, 2006, 27, 626-632.	2.5	184
45	Acetylornithine Transcarbamylase: a Novel Enzyme in Arginine Biosynthesis. Journal of Bacteriology, 2006, 188, 2974-2982.	2.2	42
46	Structure and Catalytic Mechanism of a Novel N-Succinyl-l-ornithine Transcarbamylase in Arginine Biosynthesis of Bacteroides fragilis. Journal of Biological Chemistry, 2006, 281, 20623-20631.	3.4	22
47	Long-Term Correction of Ammonia Metabolism and Prolonged Survival in Ornithine Transcarbamylase-Deficient Mice Following Liver-Directed Treatment with Adeno-associated Viral Vectors. Molecular Therapy, 2006, 14, 25-33.	8.2	76
48	Late onset N-acetylglutamate synthase deficiency caused by hypomorphic alleles. Human Mutation, 2005, 25, 293-298.	2.5	37
49	Expression, purification, crystallization and preliminary X-ray crystallographic studies of a novel acetylcitrulline deacetylase fromXanthomonas campestris. Acta Crystallographica Section F: Structural Biology Communications, 2005, 61, 676-679.	0.7	11
50	Crystal Structure of N-Acetylornithine Transcarbamylase from Xanthomonas campestris. Journal of Biological Chemistry, 2005, 280, 14366-14369.	3.4	39
51	The use of yeast mitochondria to study the properties of wild-type and mutant human mitochondrial ornithine transporter. Molecular Genetics and Metabolism, 2005, 86, 431-440.	1.1	11
52	Mammalian N-acetylglutamate synthase. Molecular Genetics and Metabolism, 2004, 81, 4-11.	1.1	39
53	Restoration of ureagenesis in N-acetylglutamate synthase deficiency by N-carbamylglutamate. Journal of Pediatrics, 2004, 145, 552-554.	1.8	58
54	Null mutations in the N-acetylglutamate synthase gene associated with acute neonatal disease and hyperammonemia. Human Genetics, 2003, 112, 364-368.	3.8	49

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55	Identification, cloning and expression of the mouse N-acetylglutamate synthase gene. Biochemical Journal, 2002, 364, 825-831.	3.7	52
56	Cloning and expression of the human N-acetylglutamate synthase gene. Biochemical and Biophysical Research Communications, 2002, 299, 581-586.	2.1	74
57	Crystal Structure of a Transcarbamylase-like Protein from the Anaerobic Bacterium Bacteroides fragilis at 2.0Ã... Resolution. Journal of Molecular Biology, 2002, 320, 899-908.	4.2	23
58	Mutations and polymorphisms in the human ornithine transcarbamylase gene. Human Mutation, 2002, 19, 93-107.	2.5	120
59	Human ornithine transcarbamylase: crystallographic insights into substrate recognition and conformational changes. Biochemical Journal, 2001, 354, 501-509.	3.7	48
60	Crystallization and preliminary X-ray crystallographic studies of wild-type human ornithine transcarbamylase and two naturally occurring mutants at position 277. Acta Crystallographica Section D: Biological Crystallography, 2001, 57, 719-721.	2.5	2
61	The clinically variable R40H mutant ornithine carbamoyltransferase shows cytosolic degradation of the precursor protein in CHO cells. Journal of Inherited Metabolic Disease, 2001, 24, 614-622.	3.6	7
62	Human ornithine transcarbamylase: crystallographic insights into substrate recognition and conformational changes. Biochemical Journal, 2001, 354, 501.	3.7	38
63	Crystal structure of human ornithine transcarbamylase complexed with carbamoyl phosphate and L-norvaline at 1.9 ? resolution. , 2000, 39, 271-277.		35
64	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. Pediatric Research, 2000, 48, 842-846.	2.3	10
65	Canine Heparan Sulfate Sulfamidase and the Molecular Pathology Underlying Sanfilippo Syndrome Type A in Dachshunds. Genomics, 2000, 68, 80-84.	2.9	25
66	Molecular Recognition by Ornithine and Aspartate Transcarbamylases. Accounts of Chemical Research, 1999, 32, 885-894.	15.6	19
67	1.85-Ã... Resolution Crystal Structure of Human Ornithine Transcarbamoylase Complexed withN-Phosphonacetyl-l-ornithine. Journal of Biological Chemistry, 1998, 273, 34247-34254.	3.4	73
68	â€˜Late Onsetâ€™ Ornithine Transcarbamylase Deficiency: Function of Three Purified Recombinant Mutant Enzymes. Human Molecular Genetics, 1997, 6, 963-968.	2.9	16
69	Expression, purification, and characterization of recombinant human glutamine synthetase. Biochemical Journal, 1997, 328, 159-163.	3.7	41
70	Expression, purification and kinetic characterization of wild-type human ornithine transcarbamylase and a recurrent mutant that produces â€˜late onsetâ€™ hyperammonaemia. Biochemical Journal, 1997, 322, 625-631.	3.7	37
71	Identification of â€˜privateâ€™ mutations in patients with ornithine transcarbamylase deficiency. Journal of Inherited Metabolic Disease, 1997, 20, 525-527.	3.6	35