Jan P Kraus

List of Publications by Year
in descending order

64 papers	2,981 citations	136950 32 h-index	175258 52 g-index
66 all docs	66 docs citations	66 times ranked	1982 citing authors

#	Article	IF	Citations
1	Cystathionine ?-synthase mutations in homocystinuria. Human Mutation, 1999, 13, 362-375.	2.5	247
2	Cystathionine Î²-Synthase: Structure, Function, Regulation, and Location of Homocystinuria-causing Mutations. Journal of Biological Chemistry, 2004, 279, 29871-29874.	3.4	204
3	Trypsin Cleavage of Human Cystathionine Î²-Synthase into an Evolutionarily Conserved Active Core: Structural and Functional Consequences. Archives of Biochemistry and Biophysics, 1998, 355, 222-232.	3.0	150
4	Regulation of Human Cystathionine Î²-Synthase byS-Adenosyl-l-methionine:Â Evidence for Two Catalytically Active Conformations Involving an Autoinhibitory Domain in the C-Terminal Regionâ€. Biochemistry, 2001, 40, 10625-10633.	2.5	150
5	Screening for mutations by expressing patient cDNA segments inE. coli: Homocystinuria due to cystathionine Î²-synthase deficiency. Human Mutation, 1992, 1, 113-123.	2.5	103
6	Structural basis of regulation and oligomerization of human cystathionine Î²-synthase, the central enzyme of transsulfuration. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, E3790-9.	7.1	89
7	Structural insight into the molecular mechanism of allosteric activation of human cystathionine Î²-synthase by S -adenosylmethionine. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, E3845-52.	7.1	86
8	Impaired Heme Binding and Aggregation of Mutant Cystathionine Î²-Synthase Subunits in Homocystinuria. American Journal of Human Genetics, 2001, 68, 1506-1513.	6.2	78
9	Human cystathionine Î²-synthase (CBS) contains two classes of binding sites for S-adenosylmethionine (SAM): complex regulation of CBS activity and stability by SAM. Biochemical Journal, 2013, 449, 109-121.	3.7	78
10	High frequency (71%) of cystathionine Î²-synthase mutation G307S in Irish homocystinuria patients. Human Mutation, 1995, 6, 177-180.	2.5	75
11	Binding of Pyridoxal 5â€˜-Phosphate to the Heme Protein Human Cystathionine Î²-Synthase. Biochemistry, 1999, 38, 2716-2724.	2.5	69
12	High homocysteine and thrombosis without connective tissue disorders are associated with a novel class of cystathionine Î²-synthase (CBS) mutations. Human Mutation, 2002, 19, 641-655.	2.5	64
13	Cystathionine Î²-synthase (human). Methods in Enzymology, 1987, 143, 388-394.	1.0	63
14	Rescue of Cystathionine Î²-Synthase (CBS) Mutants with Chemical Chaperones. Journal of Biological Chemistry, 2010, 285, 15866-15873.	3.4	63
15	Deletion Mutagenesis of Human Cystathionine Î²-Synthase. Journal of Biological Chemistry, 2002, 277, 48386-48394.	3.4	60
16	A novel transgenic mouse model of CBS-deficient homocystinuria does not incur hepatic steatosis or fibrosis and exhibits a hypercoagulative phenotype that is ameliorated by betaine treatment. Molecular Genetics and Metabolism, 2010, 101, 153-162.	1.1	60
17	Functional Properties of the Active Core of Human Cystathionine Î²-Synthase Crystals. Journal of Biological Chemistry, 2001, 276, 16-19.	3.4	58
18	Cystathionine Î³-lyase: Clinical, metabolic, genetic, and structural studies. Molecular Genetics and Metabolism, 2009, 97, 250-259.	1.1	57

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19	Cystathionine beta-synthase null homocystinuric mice fail to exhibit altered hemostasis or lowering of plasma homocysteine in response to betaine treatment. Molecular Genetics and Metabolism, 2010, 101, 163-171.	1.1	57
20	Cystathionine Î²-synthase mutations: effect of mutation topology on folding and activity. Human Mutation, 2010, 31, 809-819.	2.5	50
21	Effect of cobalt on Escherichia coli metabolism and metalloporphyrin formation. BioMetals, 2011, 24, 335-347.	4.1	50
22	Active Cystathionine Î²-Synthase Can Be Expressed in Heme-free Systems in the Presence of Metal-substituted Porphyrins or a Chemical Chaperone. Journal of Biological Chemistry, 2008, 283, 34588-34595.	3.4	48
23	Transsulfuration in Saccharomyces cerevisiae is not dependent on heme: purification and characterization of recombinant yeast cystathionine Î²-synthase. Journal of Inorganic Biochemistry, 2000, 81, 161-171.	3.5	47
24	Biogenesis of Hydrogen Sulfide and Thioethers by Cystathionine Beta-Synthase. Antioxidants and Redox Signaling, 2018, 28, 311-323.	5.4	47
25	Generation of a Hypomorphic Model of Propionic Acidemia Amenable to Gene Therapy Testing. Molecular Therapy, 2013, 21, 1316-1323.	8.2	46
26	Domain Organization, Catalysis and Regulation of Eukaryotic Cystathionine Beta-Synthases. PLoS ONE, 2014, 9, e105290.	2.5	42
27	Characterization of Four Variant Forms of Human Propionyl-CoA Carboxylase Expressed in Escherichia coli. Journal of Biological Chemistry, 2005, 280, 27719-27727.	3.4	39
28	The Heme of Cystathionine Î²-synthase Likely Undergoes a Thermally Induced Redox-Mediated Ligand Switch. Biochemistry, 2005, 44, 16785-16795.	2.5	39
29	The Redox Behavior of the Heme in Cystathionine Î²-synthase Is Sensitive to pH. Biochemistry, 2004, 43, 14684-14695.	2.5	38
30	Enzyme replacement with PEGylated cystathionine Î²-synthase ameliorates homocystinuria in murine model. Journal of Clinical Investigation, 2016, 126, 2372-2384.	8.2	37
31	Long-Term Sex-Biased Correction of Circulating Propionic Acidemia Disease Markers by Adeno-Associated Virus Vectors. Human Gene Therapy, 2015, 26, 153-160.	2.7	35
32	Ferrous Human Cystathionine Î²-Synthase Loses Activity during Enzyme Assay Due to a Ligand Switch Process. Biochemistry, 2007, 46, 13199-13210.	2.5	33
33	Purification and characterization of the wild type and truncated human cystathionine Î²-synthase enzymes expressed in E. coli. Archives of Biochemistry and Biophysics, 2008, 470, 64-72.	3.0	32
34	Targeting Cystathionine Beta-Synthase Misfolding in Homocystinuria by Small Ligands: State of the Art and Future Directions. Current Drug Targets, 2016, 17, 1455-1470.	2.1	30
35	The molecular basis of cystathionine ÃŸ-synthase (CBS) deficiency in UK and US patients with homocystinuria. Human Mutation, 2004, 23, 206-206.	2.5	28
36	Thioethers as markers of hydrogen sulfide production in homocystinurias. Biochimie, 2016, 126, 14-20.	2.6	28

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37	Potential Pharmacological Chaperones for Cystathionine Beta-Synthase-Deficient Homocystinuria. Handbook of Experimental Pharmacology, 2017, 245, 345-383.	1.8	28
38	Enzyme Replacement Therapy Ameliorates Multiple Symptoms of Murine Homocystinuria. Molecular Therapy, 2018, 26, 834-844.	8.2	28
39	Cystathionine Î²-synthase mutations in homocystinuria. Human Mutation, 1999, 13, 362.	2.5	28
40	Molecular defect in a patient with pyridoxine-responsive homocystinuria. Human Molecular Genetics, 1993, 2, 815-816.	2.9	27
41	Characterization of a cystathionine Î²-synthase allele with three mutations in cis in a patient with B6 nonresponsive homocystinuria. Human Molecular Genetics, 1994, 3, 1883-1886.	2.9	27
42	Four novel mutations in the cystathionine ?-synthase gene: Effect of a second linked mutation on the severity of the homocystinuric phenotype. , 1999, 13, 453-457.		27
43	Folding and activity of mutant cystathionine Î²-synthase depends on the position and nature of the purification tag: Characterization of the R266K CBS mutant. Protein Expression and Purification, 2012, 82, 317-324.	1.3	26
44	Coordination Chemistry of the Heme in Cystathionine Î²-Synthase: Formation of Iron(II)â€“Isonitrile Complexes. Biochemical and Biophysical Research Communications, 2001, 283, 487-492.	2.1	24
45	Conformational Properties of Nine Purified Cystathionine Î²-Synthase Mutants. Biochemistry, 2012, 51, 4755-4763.	2.5	24
46	Enzyme replacement prevents neonatal death, liver damage, and osteoporosis in murine homocystinuria. FASEB Journal, 2017, 31, 5495-5506.	0.5	24
47	Kinetic stability of cystathionine beta-synthase can be modulated by structural analogs of S-adenosylmethionine: Potential approach to pharmacological chaperone therapy for homocystinuria. Biochimie, 2016, 126, 6-13.	2.6	23
48	Marine natural products as inhibitors of cystathionine beta-synthase activity. Bioorganic and Medicinal Chemistry Letters, 2015, 25, 1064-1066.	2.2	21
49	Cobalt Cystathionine Î²-Synthase: A Cobalt-Substituted Heme Protein with a Unique Thiolate Ligation Motif. Inorganic Chemistry, 2011, 50, 4417-4427.	4.0	17
50	Mutational analysis of the cystathionine Î²-synthase gene: A splicing mutation, two missense mutations and an insertion in patients with homocystinuria. Human Mutation, 1998, 11, 332-332.	2.5	16
51	Engineering and Characterization of an Enzyme Replacement Therapy for Classical Homocystinuria. Biomacromolecules, 2017, 18, 1747-1761.	5.4	16
52	Behavior, body composition, and vascular phenotype of homocystinuric mice on methionineâ€restricted diet or enzyme replacement therapy. FASEB Journal, 2019, 33, 12477-12486.	0.5	16
53	Comparative Study of Enzyme Activity and Heme Reactivity inDrosophila melanogasterandHomo sapiensCystathionine Î²-Synthases. Biochemistry, 2013, 52, 741-751.	2.5	15
54	Import of TAT-Conjugated Propionyl Coenzyme A Carboxylase Using Models of Propionic Acidemia. Molecular and Cellular Biology, 2018, 38, .	2.3	15

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55	Detection of a novel deletion in the cystathionine Î²-synthase (CBS) gene using an improved genomic DNA based method1. FEBS Letters, 1998, 431, 175-179.	2.8	13
56	Crystal structure of cystathionine Î²-synthase from honeybee Apis mellifera. Journal of Structural Biology, 2018, 202, 82-93.	2.8	13
57	Enzyme replacement therapy prevents loss of bone and fat mass in murine homocystinuria. Human Mutation, 2018, 39, 210-218.	2.5	13
58	Purification and characterization of cystathionine Î²-synthase bearing a cobalt protoporphyrin. Archives of Biochemistry and Biophysics, 2011, 508, 25-30.	3.0	12
59	The role of surface electrostatics on the stability, function and regulation of human cystathionine Î²-synthase, a complex multidomain and oligomeric protein. Biochimica Et Biophysica Acta - Proteins and Proteomics, 2014, 1844, 1453-1462.	2.3	10
60	Oligomeric status of human cystathionine betaâ€ synthase modulates AdoMet binding. FEBS Letters, 2016, 590, 4461-4471.	2.8	8
61	Pharmacokinetics and pharmacodynamics of PEGylated truncated human cystathionine beta-synthase for treatment of homocystinuria. Life Sciences, 2018, 200, 15-25.	4.3	7
62	The Homocystinurias. , 0, , 627-650.		6
63	Mutational analysis of the cystathionine Î²â€ synthase gene: A splicing mutation, two missense mutations and an insertion in patients with homocystinuria. Human Mutation, 1998, 11, 332-332.	2.5	1
64	Purification, crystallization and preliminary crystallographic analysis of the catalytic core of cystathionine Î²-synthase fromSaccharomyces cerevisiae. Acta Crystallographica Section F, Structural Biology Communications, 2014, 70, 320-325.	0.8	0