

# Jan P Kraus

## List of Publications by Year in descending order

Source: <https://exaly.com/author-pdf/2856427/publications.pdf>

Version: 2024-02-01

64  
papers

2,981  
citations

147566

31  
h-index

189595

50  
g-index

66  
all docs

66  
docs citations

66  
times ranked

1982  
citing authors

#	ARTICLE	IF	CITATIONS
1	Cystathionine $\beta$ -synthase mutations in homocystinuria. , 1999, 13, 362-375.		247
2	Cystathionine $\beta$ -Synthase: Structure, Function, Regulation, and Location of Homocystinuria-causing Mutations. Journal of Biological Chemistry, 2004, 279, 29871-29874.	1.6	204
3	Trypsin Cleavage of Human Cystathionine $\beta$ -Synthase into an Evolutionarily Conserved Active Core: Structural and Functional Consequences. Archives of Biochemistry and Biophysics, 1998, 355, 222-232.	1.4	150
4	Regulation of Human Cystathionine $\beta$ -Synthase by S-Adenosyl-L-methionine: Evidence for Two Catalytically Active Conformations Involving an Autoinhibitory Domain in the C-Terminal Region. Biochemistry, 2001, 40, 10625-10633.	1.2	150
5	Screening for mutations by expressing patient cDNA segments in E. coli: Homocystinuria due to cystathionine $\beta$ -synthase deficiency. Human Mutation, 1992, 1, 113-123.	1.1	103
6	Structural basis of regulation and oligomerization of human cystathionine $\beta$ -synthase, the central enzyme of transsulfuration. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, E3790-9.	3.3	89
7	Structural insight into the molecular mechanism of allosteric activation of human cystathionine $\beta$ -synthase by S-adenosylmethionine. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, E3845-52.	3.3	86
8	Impaired Heme Binding and Aggregation of Mutant Cystathionine $\beta$ -Synthase Subunits in Homocystinuria. American Journal of Human Genetics, 2001, 68, 1506-1513.	2.6	78
9	Human cystathionine $\beta$ -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.	1.7	78
10	High frequency (71%) of cystathionine $\beta$ -synthase mutation G307S in Irish homocystinuria patients. Human Mutation, 1995, 6, 177-180.	1.1	75
11	Binding of Pyridoxal 5'-Phosphate to the Heme Protein Human Cystathionine $\beta$ -Synthase. Biochemistry, 1999, 38, 2716-2724.	1.2	69
12	High homocysteine and thrombosis without connective tissue disorders are associated with a novel class of cystathionine $\beta$ -synthase (CBS) mutations. Human Mutation, 2002, 19, 641-655.	1.1	64
13	Cystathionine $\beta$ -synthase (human). Methods in Enzymology, 1987, 143, 388-394.	0.4	63
14	Rescue of Cystathionine $\beta$ -Synthase (CBS) Mutants with Chemical Chaperones. Journal of Biological Chemistry, 2010, 285, 15866-15873.	1.6	63
15	Deletion Mutagenesis of Human Cystathionine $\beta$ -Synthase. Journal of Biological Chemistry, 2002, 277, 48386-48394.	1.6	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.	0.5	60
17	Functional Properties of the Active Core of Human Cystathionine $\beta$ -Synthase Crystals. Journal of Biological Chemistry, 2001, 276, 16-19.	1.6	58
18	Cystathionine $\beta$ -lyase: Clinical, metabolic, genetic, and structural studies. Molecular Genetics and Metabolism, 2009, 97, 250-259.	0.5	57

#	ARTICLE	IF	CITATIONS
19	Cystathionine beta-synthase null homocystinuric mice fail to exhibit altered hemostasis or lowering of plasma homocysteine in response to betaine treatment. <i>Molecular Genetics and Metabolism</i> , 2010, 101, 163-171.	0.5	57
20	Cystathionine $\beta$ -synthase mutations: effect of mutation topology on folding and activity. <i>Human Mutation</i> , 2010, 31, 809-819.	1.1	50
21	Effect of cobalt on <i>Escherichia coli</i> metabolism and metalloporphyrin formation. <i>BioMetals</i> , 2011, 24, 335-347.	1.8	50
22	Active Cystathionine $\beta$ -Synthase Can Be Expressed in Heme-free Systems in the Presence of Metal-substituted Porphyrins or a Chemical Chaperone. <i>Journal of Biological Chemistry</i> , 2008, 283, 34588-34595.	1.6	48
23	Transsulfuration in <i>Saccharomyces cerevisiae</i> is not dependent on heme: purification and characterization of recombinant yeast cystathionine $\beta$ -synthase. <i>Journal of Inorganic Biochemistry</i> , 2000, 81, 161-171.	1.5	47
24	Biogenesis of Hydrogen Sulfide and Thioethers by Cystathionine Beta-Synthase. <i>Antioxidants and Redox Signaling</i> , 2018, 28, 311-323.	2.5	47
25	Generation of a Hypomorphic Model of Propionic Acidemia Amenable to Gene Therapy Testing. <i>Molecular Therapy</i> , 2013, 21, 1316-1323.	3.7	46
26	Domain Organization, Catalysis and Regulation of Eukaryotic Cystathionine Beta-Synthases. <i>PLoS ONE</i> , 2014, 9, e105290.	1.1	42
27	Characterization of Four Variant Forms of Human Propionyl-CoA Carboxylase Expressed in <i>Escherichia coli</i> . <i>Journal of Biological Chemistry</i> , 2005, 280, 27719-27727.	1.6	39
28	The Heme of Cystathionine $\beta$ -synthase Likely Undergoes a Thermally Induced Redox-Mediated Ligand Switch. <i>Biochemistry</i> , 2005, 44, 16785-16795.	1.2	39
29	The Redox Behavior of the Heme in Cystathionine $\beta$ -synthase Is Sensitive to pH. <i>Biochemistry</i> , 2004, 43, 14684-14695.	1.2	38
30	Enzyme replacement with PEGylated cystathionine $\beta$ -synthase ameliorates homocystinuria in murine model. <i>Journal of Clinical Investigation</i> , 2016, 126, 2372-2384.	3.9	37
31	Long-Term Sex-Biased Correction of Circulating Propionic Acidemia Disease Markers by Adeno-Associated Virus Vectors. <i>Human Gene Therapy</i> , 2015, 26, 153-160.	1.4	35
32	Ferrous Human Cystathionine $\beta$ -Synthase Loses Activity during Enzyme Assay Due to a Ligand Switch Process. <i>Biochemistry</i> , 2007, 46, 13199-13210.	1.2	33
33	Purification and characterization of the wild type and truncated human cystathionine $\beta$ -synthase enzymes expressed in <i>E. coli</i> . <i>Archives of Biochemistry and Biophysics</i> , 2008, 470, 64-72.	1.4	32
34	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
35	The molecular basis of cystathionine $\beta$ -synthase (CBS) deficiency in UK and US patients with homocystinuria. <i>Human Mutation</i> , 2004, 23, 206-206.	1.1	28
36	Thioethers as markers of hydrogen sulfide production in homocystinurias. <i>Biochimie</i> , 2016, 126, 14-20.	1.3	28

#	ARTICLE	IF	CITATIONS
37	Potential Pharmacological Chaperones for Cystathionine Beta-Synthase-Deficient Homocystinuria. Handbook of Experimental Pharmacology, 2017, 245, 345-383.	0.9	28
38	Enzyme Replacement Therapy Ameliorates Multiple Symptoms of Murine Homocystinuria. Molecular Therapy, 2018, 26, 834-844.	3.7	28
39	Cystathionine $\beta$ -synthase mutations in homocystinuria. Human Mutation, 1999, 13, 362.	1.1	28
40	Molecular defect in a patient with pyridoxine-responsive homocystinuria. Human Molecular Genetics, 1993, 2, 815-816.	1.4	27
41	Characterization of a cystathionine $\beta$ -synthase allele with three mutations in cis in a patient with B6 nonresponsive homocystinuria. Human Molecular Genetics, 1994, 3, 1883-1886.	1.4	27
42	Four novel mutations in the cystathionine $\beta$ -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 $\beta$ -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.	0.6	26
44	Coordination Chemistry of the Heme in Cystathionine $\beta$ -Synthase: Formation of Iron(II) $\pi$ -Isonitrile Complexes. Biochemical and Biophysical Research Communications, 2001, 283, 487-492.	1.0	24
45	Conformational Properties of Nine Purified Cystathionine $\beta$ -Synthase Mutants. Biochemistry, 2012, 51, 4755-4763.	1.2	24
46	Enzyme replacement prevents neonatal death, liver damage, and osteoporosis in murine homocystinuria. FASEB Journal, 2017, 31, 5495-5506.	0.2	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.	1.3	23
48	Marine natural products as inhibitors of cystathionine beta-synthase activity. Bioorganic and Medicinal Chemistry Letters, 2015, 25, 1064-1066.	1.0	21
49	Cobalt Cystathionine $\beta$ -Synthase: A Cobalt-Substituted Heme Protein with a Unique Thiolate Ligation Motif. Inorganic Chemistry, 2011, 50, 4417-4427.	1.9	17
50	Mutational analysis of the cystathionine $\beta$ -synthase gene: A splicing mutation, two missense mutations and an insertion in patients with homocystinuria. Human Mutation, 1998, 11, 332-332.	1.1	16
51	Engineering and Characterization of an Enzyme Replacement Therapy for Classical Homocystinuria. Biomacromolecules, 2017, 18, 1747-1761.	2.6	16
52	Behavior, body composition, and vascular phenotype of homocystinuric mice on methionine-restricted diet or enzyme replacement therapy. FASEB Journal, 2019, 33, 12477-12486.	0.2	16
53	Comparative Study of Enzyme Activity and Heme Reactivity in <i>Drosophila melanogaster</i> and <i>Homo sapiens</i> Cystathionine $\beta$ -Synthases. Biochemistry, 2013, 52, 741-751.	1.2	15
54	Import of TAT-Conjugated Propionyl Coenzyme A Carboxylase Using Models of Propionic Acidemia. Molecular and Cellular Biology, 2018, 38, .	1.1	15

#	ARTICLE	IF	CITATIONS
55	Detection of a novel deletion in the cystathionine $\beta$ -synthase (CBS) gene using an improved genomic DNA based method. FEBS Letters, 1998, 431, 175-179.	1.3	13
56	Crystal structure of cystathionine $\beta$ -synthase from honeybee <i>Apis mellifera</i> . Journal of Structural Biology, 2018, 202, 82-93.	1.3	13
57	Enzyme replacement therapy prevents loss of bone and fat mass in murine homocystinuria. Human Mutation, 2018, 39, 210-218.	1.1	13
58	Purification and characterization of cystathionine $\beta$ -synthase bearing a cobalt protoporphyrin. Archives of Biochemistry and Biophysics, 2011, 508, 25-30.	1.4	12
59	The role of surface electrostatics on the stability, function and regulation of human cystathionine $\beta$ -synthase, a complex multidomain and oligomeric protein. Biochimica Et Biophysica Acta - Proteins and Proteomics, 2014, 1844, 1453-1462.	1.1	10
60	Oligomeric status of human cystathionine $\beta$ -synthase modulates AdoMet binding. FEBS Letters, 2016, 590, 4461-4471.	1.3	8
61	Pharmacokinetics and pharmacodynamics of PEGylated truncated human cystathionine $\beta$ -synthase for treatment of homocystinuria. Life Sciences, 2018, 200, 15-25.	2.0	7
62	The Homocystinurias. , 0, , 627-650.		6
63	Mutational analysis of the cystathionine $\beta$ -synthase gene: A splicing mutation, two missense mutations and an insertion in patients with homocystinuria. Human Mutation, 1998, 11, 332-332.	1.1	1
64	Purification, crystallization and preliminary crystallographic analysis of the catalytic core of cystathionine $\beta$ -synthase from <i>Saccharomyces cerevisiae</i> . Acta Crystallographica Section F, Structural Biology Communications, 2014, 70, 320-325.	0.4	0