Grazia Isaya

List of Publications by Year
in descending order

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67 papers	4,352 citations	94433 37 h-index	106344 65 g-index
67 all docs	67 docs citations	67 times ranked	3130 citing authors

#	Article	IF	Citations
1	Frataxin Acts as an Iron Chaperone Protein to Modulate Mitochondrial Aconitase Activity. Science, 2004, 305, 242-245.	12.6	361
2	Iron-Dependent Self-Assembly of Recombinant Yeast Frataxin: Implications for Friedreich Ataxia. American Journal of Human Genetics, 2000, 67, 549-562.	6.2	248
3	Assembly and iron-binding properties of human frataxin, the protein deficient in Friedreich ataxia. Human Molecular Genetics, 2002, 11, 217-227.	2.9	180
4	Mitochondrial iron detoxification is a primary function of frataxin that limits oxidative damage and preserves cell longevity. Human Molecular Genetics, 2006, 15, 467-479.	2.9	179
5	Yeast Frataxin Sequentially Chaperones and Stores Iron by Coupling Protein Assembly with Iron Oxidation. Journal of Biological Chemistry, 2003, 278, 31340-31351.	3.4	145
6	Human frataxin maintains mitochondrial iron homeostasis in Saccharomyces cerevisiae. Human Molecular Genetics, 2000, 9, 2523-2530.	2.9	140
7	Cleavage of precursors by the mitochondrial processing peptidase requires a compatible mature protein or an intermediate octapeptide.. Journal of Cell Biology, 1991, 113, 65-76.	5.2	129
8	Friedreich Ataxia: From GAA Tripletâ€“Repeat Expansion to Frataxin Deficiency. American Journal of Human Genetics, 2001, 69, 15-24.	6.2	126
9	Physical Evidence that Yeast Frataxin Is an Iron Storage Proteinâ€. Biochemistry, 2002, 41, 6798-6804.	2.5	120
10	Ironâ€“sulfur cluster synthesis, iron homeostasis and oxidative stress in Friedreich ataxia. Molecular and Cellular Neurosciences, 2013, 55, 50-61.	2.2	109
11	Mistargeting of peroxisomal L-alanine:glyoxylate aminotransferase to mitochondria in primary hyperoxaluria patients depends upon activation of a cryptic mitochondrial targeting sequence by a point mutation.. Proceedings of the National Academy of Sciences of the United States of America, 1991, 88, 10900-10904.	7.1	108
12	Cryptic proteolytic activity of dihydrolipoamide dehydrogenase. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 6158-6163.	7.1	107
13	Two-step Processing of Human Frataxin by Mitochondrial Processing Peptidase. Journal of Biological Chemistry, 2000, 275, 41469-41475.	3.4	101
14	The expression of human mitochondrial ferritin rescues respiratory function infrataxin-deficient yeast. Human Molecular Genetics, 2004, 13, 2279-2288.	2.9	100
15	Rat liver mitochondrial intermediate peptidase (MIP): purification and initial characterization.. EMBO Journal, 1992, 11, 2803-2809.	7.8	99
16	Yeast and Human Frataxin Are Processed to Mature Form in Two Sequential Steps by the Mitochondrial Processing Peptidase. Journal of Biological Chemistry, 1999, 274, 22763-22769.	3.4	99
17	Prediction and Identification of New Natural Substrates of the Yeast Mitochondrial Intermediate Peptidase. Journal of Biological Chemistry, 1995, 270, 27366-27373.	3.4	98
18	Assembly of Human Frataxin Is a Mechanism for Detoxifying Redox-Active Iron. Biochemistry, 2005, 44, 537-545.	2.5	95

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19	Chelatases: distort to select?. Trends in Biochemical Sciences, 2006, 31, 135-142.	7.5	94
20	Reduction in frataxin causes progressive accumulation of mitochondrial damage. Human Molecular Genetics, 2003, 12, 3331-3342.	2.9	91
21	Human Acyl-CoA Dehydrogenase-9 Plays a Novel Role in the Mitochondrial Î²-Oxidation of Unsaturated Fatty Acids. Journal of Biological Chemistry, 2005, 280, 32309-32316.	3.4	88
22	Mitochondrial iron-sulfur cluster dysfunction in neurodegenerative disease. Frontiers in Pharmacology, 2014, 5, 29.	3.5	82
23	The Ferroxidase Activity of Yeast Frataxin. Journal of Biological Chemistry, 2002, 277, 38589-38595.	3.4	80
24	The Structures of Frataxin Oligomers Reveal the Mechanism for the Delivery and Detoxification of Iron. Structure, 2006, 14, 1535-1546.	3.3	78
25	MIP1, a new yeast gene homologous to the rat mitochondrial intermediate peptidase gene, is required for oxidative metabolism in Saccharomyces cerevisiae.. Molecular and Cellular Biology, 1994, 14, 5603-5616.	2.3	75
26	Structure of Frataxin Iron Cores:Â An X-ray Absorption Spectroscopic Studyâ€. Biochemistry, 2003, 42, 5971-5976.	2.5	68
27	Lateral-flow immunoassay for the frataxin protein in Friedreichâ€™s ataxia patients and carriers. Molecular Genetics and Metabolism, 2008, 94, 491-497.	1.1	67
28	Normal and Friedreich Ataxia Cells Express Different Isoforms of Frataxin with Complementary Roles in Iron-Sulfur Cluster Assembly. Journal of Biological Chemistry, 2010, 285, 38486-38501.	3.4	67
29	Advancements in the pathophysiology of Friedreichâ€™s Ataxia and new prospects for treatments. Molecular Genetics and Metabolism, 2007, 92, 23-35.	1.1	65
30	Mitochondrial intermediate peptidase and the yeast frataxin homolog together maintain mitochondrial iron homeostasis in Saccharomyces cerevisiae. Human Molecular Genetics, 1999, 8, 1099-1110.	2.9	60
31	PMPCAmutations cause abnormal mitochondrial protein processing in patients with non-progressive cerebellar ataxia. Brain, 2015, 138, 1505-1517.	7.6	58
32	Oligomeric Yeast Frataxin Drives Assembly of Core Machinery for Mitochondrial Iron-Sulfur Cluster Synthesis. Journal of Biological Chemistry, 2009, 284, 21971-21980.	3.4	53
33	Amino-terminal octapeptides function as recognition signals for the mitochondrial intermediate peptidase. Journal of Biological Chemistry, 1992, 267, 7904-10.	3.4	53
34	Supramolecular Assemblies of Human Frataxin are Formed via Subunitâ€“Subunit Interactions Mediated by a Non-conserved Amino-terminal Region. Journal of Molecular Biology, 2005, 345, 433-439.	4.2	52
35	Sequence analysis of rat mitochondrial intermediate peptidase: similarity to zinc metallopeptidases and to a putative yeast homologue.. Proceedings of the National Academy of Sciences of the United States of America, 1992, 89, 8317-8321.	7.1	47
36	Structural Basis of the Iron Storage Function of Frataxin from Single-Particle Reconstruction of the Iron-Loaded Oligomer. Biochemistry, 2008, 47, 4948-4954.	2.5	40

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37	Mutations in the Dimer Interface of Dihydrolipoamide Dehydrogenase Promote Site-specific Oxidative Damages in Yeast and Human Cells. Journal of Biological Chemistry, 2011, 286, 40232-40245.	3.4	40
38	Mammalian Mitochondrial Intermediate Peptidase: Structure/Function Analysis of a New Homologue from Schizophyllum commune and Relationship to Thimet Oligopeptidases. Genomics, 1995, 28, 450-461.	2.9	33
39	Assembly of the Iron-binding Protein Frataxin in Saccharomyces cerevisiae Responds to Dynamic Changes in Mitochondrial Iron Influx and Stress Level. Journal of Biological Chemistry, 2008, 283, 31500-31510.	3.4	33
40	Cloning, Expression, and Chromosomal Assignment of the Human Mitochondrial Intermediate Peptidase Gene (MIPEP). Genomics, 1997, 40, 493-496.	2.9	31
41	Mitochondrial import and processing of mutant human ornithine transcarbamylase precursors in cultured cells.. Molecular and Cellular Biology, 1988, 8, 5150-5158.	2.3	30
42	Functional studies of frataxin. Acta Paediatrica, International Journal of Paediatrics, 2004, 93, 68-71.	1.5	27
43	[33] Mitochondrial intermediate peptidase. Methods in Enzymology, 1995, 248, 556-567.	1.0	25
44	Functional and Genomic Analysis of the Human Mitochondrial Intermediate Peptidase, a Putative Protein Partner of Frataxin. Genomics, 2000, 65, 104-112.	2.9	25
45	Missense Mutations Linked to Friedreich Ataxia Have Different but Synergistic Effects on Mitochondrial Frataxin Isoforms. Journal of Biological Chemistry, 2013, 288, 4116-4127.	3.4	25
46	Partial conservation of functions between eukaryotic frataxin and theEscherichia colifrataxin homolog CyaY. FEMS Yeast Research, 2007, 7, 1276-1284.	2.3	24
47	Architecture of the Human Mitochondrial Iron-Sulfur Cluster Assembly Machinery. Journal of Biological Chemistry, 2016, 291, 21296-21321.	3.4	24
48	MIP1, a New Yeast Gene Homologous to the Rat Mitochondrial Intermediate Peptidase Gene, Is Required for Oxidative Metabolism in Saccharomyces cerevisiae. Molecular and Cellular Biology, 1994, 14, 5603-5616.	2.3	24
49	A Comparative Effectiveness Study of Newborn Screening Methods for Four Lysosomal Storage Disorders. International Journal of Neonatal Screening, 2020, 6, 44.	3.2	23
50	The Structure of the Complex between Yeast Frataxin and Ferrochelatase. Journal of Biological Chemistry, 2016, 291, 11887-11898.	3.4	22
51	Mutations in a Putative Zinc-Binding Domain Inactivate the Mitochondrial Intermediate Peptidase. Biochemical and Biophysical Research Communications, 1996, 226, 822-829.	2.1	21
52	Mammalian Pitrilysin: Substrate Specificity and Mitochondrial Targeting. Biochemistry, 2009, 48, 2868-2877.	2.5	21
53	Oligomerization Propensity and Flexibility of Yeast Frataxin Studied by X-ray Crystallography and Small-Angle X-ray Scattering. Journal of Molecular Biology, 2011, 414, 783-797.	4.2	21
54	The Molecular Basis of Iron-induced Oligomerization of Frataxin and the Role of the Ferroxidation Reaction in Oligomerization. Journal of Biological Chemistry, 2013, 288, 8156-8167.	3.4	21

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55	Molecular and functional characterization of a human frataxin mutation found in hypertrophic cardiomyopathy. Molecular Genetics and Metabolism, 2005, 85, 280-285.	1.1	20
56	Architecture of the Yeast Mitochondrial Iron-Sulfur Cluster Assembly Machinery. Journal of Biological Chemistry, 2016, 291, 10378-10398.	3.4	17
57	Protein import and processing reconstituted with isolated rat liver mitochondria and recombinant mitochondrial processing peptidase. Methods, 2002, 26, 298-306.	3.8	16
58	Iron-induced oligomerization of human FXN81-210 and bacterial CyaY frataxin and the effect of iron chelators. PLoS ONE, 2017, 12, e0188937.	2.5	16
59	Mitochondrial Import and Processing of Mutant Human Ornithine Transcarbamylase Precursors in Cultured Cells. Molecular and Cellular Biology, 1988, 8, 5150-5158.	2.3	15
60	Ironing out a therapy for Friedreich ataxia. Blood, 2007, 110, 1-2.	1.4	11
61	High-Throughput Immunoassay for the Biochemical Diagnosis of Friedreich Ataxia in Dried Blood Spots and Whole Blood. Clinical Chemistry, 2013, 59, 1461-1469.	3.2	10
62	Zinc and the iron donor frataxin regulate oligomerization of the scaffold protein to form new Feâ€“S cluster assembly centers. Metallomics, 2017, 9, 773-801.	2.4	6
63	Reply: Autosomal recessive cerebellar ataxia caused by a homozygous mutation inPMPCA. Brain, 2016, 139, e20-e20.	7.6	3
64	Mitochondrial intermediate peptidase. , 2004, , 366-369.		3
65	Defining the Architecture of the Core Machinery for the Assembly of Feâ€“S Clusters in Human Mitochondria. Methods in Enzymology, 2017, 595, 107-160.	1.0	2
66	SAXS and stability studies of iron-induced oligomers of bacterial frataxin CyaY. PLoS ONE, 2017, 12, e0184961.	2.5	1
67	Frataxin degrading peptidase: A multifunctional regulator of mitochondrial energy production and iron balance. FASEB Journal, 2006, 20, A48.	0.5	0