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	A Comparative Effectiveness Study of Newborn Screening Methods for Four Lysosomal Storage Disorders. International Journal of Neonatal Screening, 2020, 6, 44.	3.2	23
2	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
3	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
4	SAXS and stability studies of iron-induced oligomers of bacterial frataxin CyaY. PLoS ONE, 2017, 12, e0184961.	2.5	1
5	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
6	The Structure of the Complex between Yeast Frataxin and Ferrochelatase. Journal of Biological Chemistry, 2016, 291, 11887-11898.	3.4	22
7	Architecture of the Human Mitochondrial Iron-Sulfur Cluster Assembly Machinery. Journal of Biological Chemistry, 2016, 291, 21296-21321.	3.4	24
8	Reply: Autosomal recessive cerebellar ataxia caused by a homozygous mutation inPMPCA. Brain, 2016, 139, e20-e20.	7.6	3
9	Architecture of the Yeast Mitochondrial Iron-Sulfur Cluster Assembly Machinery. Journal of Biological Chemistry, 2016, 291, 10378-10398.	3.4	17
10	PMPCAmutations cause abnormal mitochondrial protein processing in patients with non-progressive cerebellar ataxia. Brain, 2015, 138, 1505-1517.	7.6	58
11	Mitochondrial iron-sulfur cluster dysfunction in neurodegenerative disease. Frontiers in Pharmacology, 2014, 5, 29.	3.5	82
12	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
13	Ironâ€“sulfur cluster synthesis, iron homeostasis and oxidative stress in Friedreich ataxia. Molecular and Cellular Neurosciences, 2013, 55, 50-61.	2.2	109
14	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
15	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
16	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
17	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
18	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

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19	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
20	Mammalian Pitrilysin: Substrate Specificity and Mitochondrial Targeting. Biochemistry, 2009, 48, 2868-2877.	2.5	21
21	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
22	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
23	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
24	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
25	Ironing out a therapy for Friedreich ataxia. Blood, 2007, 110, 1-2.	1.4	11
26	Advancements in the pathophysiology of Friedreichâ€™s Ataxia and new prospects for treatments. Molecular Genetics and Metabolism, 2007, 92, 23-35.	1.1	65
27	Partial conservation of functions between eukaryotic frataxin and theEscherichia colifrataxin homolog CyaY. FEMS Yeast Research, 2007, 7, 1276-1284.	2.3	24
28	The Structures of Frataxin Oligomers Reveal the Mechanism for the Delivery and Detoxification of Iron. Structure, 2006, 14, 1535-1546.	3.3	78
29	Chelatases: distort to select?. Trends in Biochemical Sciences, 2006, 31, 135-142.	7.5	94
30	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
31	Frataxin degrading peptidase: A multifunctional regulator of mitochondrial energy production and iron balance. FASEB Journal, 2006, 20, A48.	0.5	0
32	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
33	Assembly of Human Frataxin Is a Mechanism for Detoxifying Redox-Active Iron. Biochemistry, 2005, 44, 537-545.	2.5	95
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	Molecular and functional characterization of a human frataxin mutation found in hypertrophic cardiomyopathy. Molecular Genetics and Metabolism, 2005, 85, 280-285.	1.1	20
36	Frataxin Acts as an Iron Chaperone Protein to Modulate Mitochondrial Aconitase Activity. Science, 2004, 305, 242-245.	12.6	361

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37	The expression of human mitochondrial ferritin rescues respiratory function in frataxin-deficient yeast. Human Molecular Genetics, 2004, 13, 2279-2288.	2.9	100
38	Functional studies of frataxin. Acta Paediatrica, International Journal of Paediatrics, 2004, 93, 68-71.	1.5	27
39	Mitochondrial intermediate peptidase. , 2004, , 366-369.		3
40	Structure of Frataxin Iron Cores:Â An X-ray Absorption Spectroscopic Studyâ€. Biochemistry, 2003, 42, 5971-5976.	2.5	68
41	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
42	Reduction in frataxin causes progressive accumulation of mitochondrial damage. Human Molecular Genetics, 2003, 12, 3331-3342.	2.9	91
43	The Ferroxidase Activity of Yeast Frataxin. Journal of Biological Chemistry, 2002, 277, 38589-38595.	3.4	80
44	Assembly and iron-binding properties of human frataxin, the protein deficient in Friedreich ataxia. Human Molecular Genetics, 2002, 11, 217-227.	2.9	180
45	Physical Evidence that Yeast Frataxin Is an Iron Storage Proteinâ€. Biochemistry, 2002, 41, 6798-6804.	2.5	120
46	Protein import and processing reconstituted with isolated rat liver mitochondria and recombinant mitochondrial processing peptidase. Methods, 2002, 26, 298-306.	3.8	16
47	Friedreich Ataxia: From GAA Tripletâ€“Repeat Expansion to Frataxin Deficiency. American Journal of Human Genetics, 2001, 69, 15-24.	6.2	126
48	Two-step Processing of Human Frataxin by Mitochondrial Processing Peptidase. Journal of Biological Chemistry, 2000, 275, 41469-41475.	3.4	101
49	Human frataxin maintains mitochondrial iron homeostasis in Saccharomyces cerevisiae. Human Molecular Genetics, 2000, 9, 2523-2530.	2.9	140
50	Functional and Genomic Analysis of the Human Mitochondrial Intermediate Peptidase, a Putative Protein Partner of Frataxin. Genomics, 2000, 65, 104-112.	2.9	25
51	Iron-Dependent Self-Assembly of Recombinant Yeast Frataxin: Implications for Friedreich Ataxia. American Journal of Human Genetics, 2000, 67, 549-562.	6.2	248
52	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
53	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
54	Cloning, Expression, and Chromosomal Assignment of the Human Mitochondrial Intermediate Peptidase Gene (MIPEP). Genomics, 1997, 40, 493-496.	2.9	31

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55	Mutations in a Putative Zinc-Binding Domain Inactivate the Mitochondrial Intermediate Peptidase. Biochemical and Biophysical Research Communications, 1996, 226, 822-829.	2.1	21
56	[33] Mitochondrial intermediate peptidase. Methods in Enzymology, 1995, 248, 556-567.	1.0	25
57	Prediction and Identification of New Natural Substrates of the Yeast Mitochondrial Intermediate Peptidase. Journal of Biological Chemistry, 1995, 270, 27366-27373.	3.4	98
58	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
59	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
60	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
61	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
62	Rat liver mitochondrial intermediate peptidase (MIP): purification and initial characterization.. EMBO Journal, 1992, 11, 2803-2809.	7.8	99
63	Amino-terminal octapeptides function as recognition signals for the mitochondrial intermediate peptidase. Journal of Biological Chemistry, 1992, 267, 7904-10.	3.4	53
64	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
65	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
66	Mitochondrial import and processing of mutant human ornithine transcarbamylase precursors in cultured cells.. Molecular and Cellular Biology, 1988, 8, 5150-5158.	2.3	30
67	Mitochondrial Import and Processing of Mutant Human Ornithine Transcarbamylase Precursors in Cultured Cells. Molecular and Cellular Biology, 1988, 8, 5150-5158.	2.3	15