

ANGEL LUIS PEY RODRÍGUEZ

Grupo de Investigación: **BIOMOLECULAS** (Cod.: BIO223)

Departamento: Universidad de Granada. Química Física

Citas en Google Scholar: <https://scholar.google.es/citations?user=b0qdE4YAAAAJ&hl=es>

Código ORCID: <http://orcid.org/0000-0001-7706-3243>

RG: https://www.researchgate.net/profile/Angel_Pey

Mendeley: <https://www.mendeley.com/profiles/angel-pey/>

Correo electrónico: angelpey@ugr.es

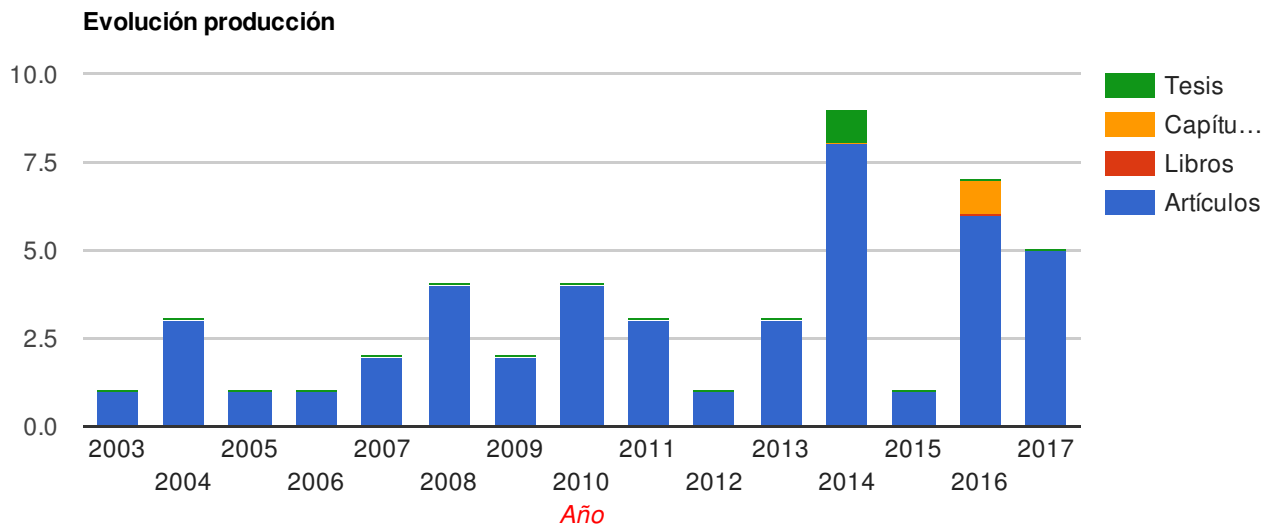
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Ficha del Directorio

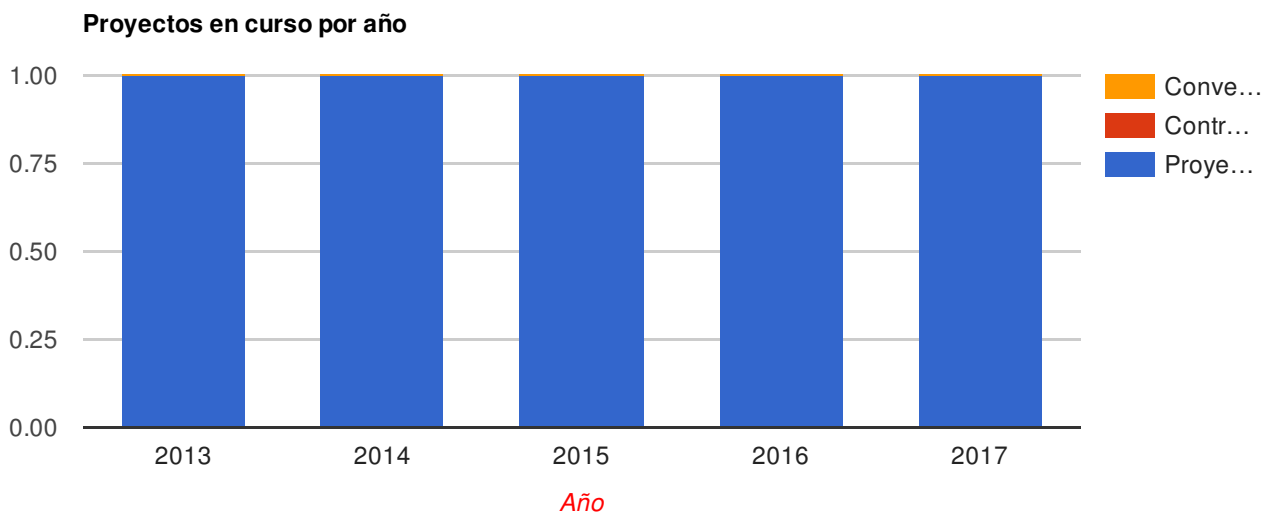
Producción 47

Artículos (45) Libros (0) Capítulos de Libros (1) Tesis dirigidas (1)



Proyectos dirigidos 1

Proyectos (1) Contratos (0) Convenios (0)



Actividades 0

Titulo publicación	Fuente	Tipo	Fecha
A mechanism for cancer-associated inactivation of nqo1 due to p187s and its reactivation by the consensus mutation h80r	Febs letters	Articulo	2017
Enhanced vulnerability of human proteins towards disease-associated inactivation through divergent evolution	Human molecular genetics	Articulo	2017
Intrinsically disordered chromatin protein nupr1 binds to the c-terminal region of polycomb ring1b	Proceedings of the national academy of sciences of the united states of america	Articulo	2017
Site-to-site interdomain communication may mediate different loss-of-function mechanisms in a cancer-associated nqo1 polymorphism.	Scientific reports	Articulo	2017
Structural basis of the oncogenic interaction of phosphatase prl-1 with the magnesium transporter cnm2.	Journal of biological chemistry	Articulo	2017
Caenorhabditis elegans alanine:glyoxylate aminotransferase is a mitochondrial and cold-adapted ortholog of peroxisomal human agt1 key to understand between-species divergence in glyoxylate metabolism	Biochimica et biophysica acta. proteins and proteomics	Articulo	2016
Calorimetric approaches to studying complex protein structure-function stability relationships in conformational diseases: the case of cystathionine β -synthase.	Biocalorimetry: foundations and contemporary approaches	Capítulo de libro	2016
Conformational dynamics is key to understanding loss-of-function of nqo1 cancer-associated polymorphisms and its correction by pharmacological ligands.	Scientific reports	Articulo	2016
Experimental and computational evidence on conformational fluctuations as a source of catalytic defects in genetic diseases	Rsc advances: an international journal to further the chemical sciences	Articulo	2016
Kinetic stability of cystathionine beta-synthase can be modulated by structural analogs of s-adenosylmethionine: potential approach to pharmacological chaperone therapy for homocystinuria.	Biochimie	Articulo	2016
Molecular basis of classic galactosemia from the structure of human galactose 1-phosphate uridylyltransferase	Human molecular genetics	Articulo	2016
The chondroitin sulfate/dermatan sulfate 4-o-endosulfatase from marine bacterium vibrio sp fc509 is a dimeric species: biophysical characterization of an endosulfatase.	Biochimie	Articulo	2016
Molecular recognition of pts-1 cargo proteins by pex5p: implications for protein mistargeting in primary hyperoxaluria	Biomolecules	Articulo	2015
Domain organization, catalysis and regulation of eukaryotic cystathionine beta-synthases	Plos one	Articulo	2014
Fad binding overcomes defects in activity and stability displayed by cancer-associated variants of human nqo1	Biochimica et biophysica acta. molecular basis of disease	Articulo	2014
Insights into human phosphoglycerate kinase 1 deficiency as a conformational disease from biochemical, biophysical, and in vitro expression analyses	Journal of inherited metabolic disease	Articulo	2014
Molecular mechanisms underlying primary hyperoxaluria type i and new therapeutic approaches	Universidad de granada	Tesis doctoral	2014
The consensus-based approach for gene/enzyme replacement therapies and crystallization strategies: the case of human alanine:glyoxylate aminotransferase	Biochemical journal	Articulo	2014
The lower limits for protein stability and foldability in primary hyperoxaluria type i	Biochimica et biophysica acta. proteins and proteomics	Articulo	2014
The metastability of human udp-galactose 4-epimerase (gale) is	Archives of biochemistry		

increased by variants associated with type iii galactosemia but decreased by substrate and cofactor binding	Archives of biochemistry and biophysics	Articulo	2014
The role of surface electrostatics on the stability, function and regulation of human cystathionine β -synthase, a complex multidomain and oligomeric protein	Biochimica et biophysica acta. proteins and proteomics	Articulo	2014
Ph-dependent relationship between thermodynamic and kinetic stability in the denaturation of human phosphoglycerate kinase 1	Biochimie	Articulo	2014
Structural and energetic basis of protein kinetic destabilization in human phosphoglycerate kinase 1 deficiency.	Biochemistry (easton)	Articulo	2013
The interplay between protein stability and dynamics in conformational diseases: the case of hpgk1 deficiency.	Biochimica et biophysica acta. proteins and proteomics	Articulo	2013
The role of protein denaturation energetics and molecular chaperones in the aggregation and mistargeting of mutants causing primary hyperoxaluria type i	Plos one	Articulo	2013
Conformational properties of nine purified cystathionine β -synthase mutants	Biochemistry	Articulo	2012
Divergence in enzyme regulation between c. elegans and human tyrosine hydroxylase, the key enzyme in the synthesis of dopamine.	Biochemical journal	Articulo	2011
Role of low native state kinetic stability and interaction of partially unfolded states with molecular chaperones in the mitochondrial protein mistargeting associated with primary hyperoxaluria	Amino acids	Articulo	2011
The regulatory subunit of pka-i remains partially structured and undergoes ζ -aggregation upon thermal denaturation	Plos one	Articulo	2011
Effect of pharmacological chaperones on brain tyrosine hydroxylase and tryptophan hydroxylase 2.	Journal of neurochemistry	Articulo	2010
Modulation of buried ionizable groups in proteins with engineered surface charge	Journal of physics and chemistry of solids	Articulo	2010
Phenylalanine hydroxylase expression in primary rat hepatocytes is modulated by oxygen concentration	Molecular genetics and metabolism	Articulo	2010
Superstoichiometric binding of l-phe to phenylalanine hydroxylase from caenorhabditis elegans: evolutionary implications	Amino acids	Articulo	2010
Biochemical characterization of mutant phenylalanine hydroxylase enzymes and correlation with clinical presentation in hyperphenylalaninaemic patients	Journal of inherited metabolic disease	Articulo	2009
Iron binding effects on the kinetic stability and unfolding energetics of a thermophilic phenylalanine hydroxylase from chloroflexus aurantiacus	Journal of biological inorganic chemistry	Articulo	2009
Anabolic function of phenylalanine hydroxylase in caenorhabditis elegans	The faseb journal	Articulo	2008
Engineering proteins with tunable thermodynamic and kinetic stabilities	Proteins: structure, function, and bioinformatics	Articulo	2008
Identification of pharmacological chaperones as potential therapeutic agents to treat phenylketonuria	Journal of clinical investigation	Articulo	2008
Thermoplasma acidophilum cdc6 protein stimulates mcm helicase activity by regulation its atpase activity	Nucleic acids research	Articulo	2008
Predicted effects of missense mutations of native-state stability account for phenotypic outcome in phenylketonuria, a paradigm of misfolding diseases	American journal of human genetics	Articulo	2007
Structure of phenylalanine hydroxylase from colwellia psychrerythraea 34h, a monomeric cold active enzyme with local flexibility around the active site and high overall stability	Journal of biological chemistry	Articulo	2007

Specific interaction of the diastereomers 7(r)- and 7(s)-tetrahydrobiopterin with phenylalanine hydroxylase: implications for understanding primapterinuria and vitiligo	The faseb journal	Articulo	2006
The activity of wild type and mutant phenylalanine hydroxylase and its regulation by phenylalanine and tetrahydrobiopterin at physiological and pathological concentrations: and isothermal titration calorimetry study	Molecular genetics and metabolism	Articulo	2005
Correction of kinetic and stability defects by tetrahydrobiopterin in phenylketonuria patients with certain phenylalanine hydroxylase mutations	Proceedings of the national academy of sciences of the united states of america	Articulo	2004
Mechanisms underlying responsiveness to tetrahydrobiopterin in mild phenylketonuria mutations	Human mutation	Articulo	2004
Thermodynamic characterization of the binding of tetrahydropterins to phenylalanine hydroxylase	Journal of physics and chemistry of solids	Articulo	2004
Phenylketonuria: genotype-phenotype correlations based on expression analysis of structural and functional mutations in pah	Human mutation	Articulo	2003

	Título proyecto	Tipo	Inicio	Fin
1	Nuevas estrategias terapéuticas para el tratamiento de enfermedades genéticas causadas por alteraciones en el plegamiento y el tráfico intracelular de proteínas	Proyecto	6/27/13	9/1/17

Actividades 0

Título actividad	Fuente	Tipo	Fecha
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Colaboradores

- NOEL MESA TORRES (9)
- ENCARNACIÓN MEDINA CARMONA (5)
- JOSE MANUEL SANCHEZ RUIZ (4)
- DAVID RODRIGUEZ LARREA (2)
- JOSE ANTONIO GAVIRA GALLARDO (2)
- ROGELIO JESÚS PALOMINO MORALES (2)
- BERTRAND MOREL (1)
- MARÍA DEL MAR GARCÍA MIRA (1)
- MIGUEL BURGOS POYATOS (1)
- RAQUEL GODOY RUIZ (1)