Raul Estevez

List of Publications by Year
in descending order

72 papers	5,332 citations	172457 29 h-index	95266 68 g-index
77 all docs	77 docs citations	77 times ranked	4007 citing authors

#	Article	IF	Citations
1	Molecular Biology of Mammalian Plasma Membrane Amino Acid Transporters. Physiological Reviews, 1998, 78, 969-1054.	28.8	778
2	Barttin is a Cl- channel Î²-subunit crucial for renal Cl- reabsorption and inner ear K+ secretion. Nature, 2001, 414, 558-561.	27.8	538
3	Identification of a Membrane Protein, LAT-2, That Co-expresses with 4F2 Heavy Chain, an L-type Amino Acid Transport Activity with Broad Specificity for Small and Large Zwitterionic Amino Acids. Journal of Biological Chemistry, 1999, 274, 19738-19744.	3.4	356
4	Identification and Characterization of a Membrane Protein (y+L Amino Acid Transporter-1) That Associates with 4F2hc to Encode the Amino Acid Transport Activity y+L. Journal of Biological Chemistry, 1998, 273, 32437-32445.	3.4	304
5	Identification of SLC7A7, encoding y+LAT-1, as the lysinuric protein intolerance gene. Nature Genetics, 1999, 21, 293-296.	21.4	286
6	Non-type I cystinuria caused by mutations in SLC7A9, encoding a subunit (bo,+AT) of rBAT. Nature Genetics, 1999, 23, 52-57.	21.4	280
7	Identification of LAT4, a Novel Amino Acid Transporter with System L Activity. Journal of Biological Chemistry, 2005, 280, 12002-12011.	3.4	216
8	Conservation of Chloride Channel Structure Revealed by an Inhibitor Binding Site in ClC-1. Neuron, 2003, 38, 47-59.	8.1	161
9	Obligatory Amino Acid Exchange via Systems bo,+-like and y+L-like. Journal of Biological Chemistry, 1996, 271, 17761-17770.	3.4	158
10	Mutant GlialCAM Causes Megalencephalic Leukoencephalopathy with Subcortical Cysts, Benign Familial Macrocephaly, and Macrocephaly with Retardation and Autism. American Journal of Human Genetics, 2011, 88, 422-432.	6.2	148
11	Functional and structural conservation of CBS domains from CLC chloride channels. Journal of Physiology, 2004, 557, 363-378.	2.9	131
12	Megalencephalic leukoencephalopathy with subcortical cysts: chronic white matter oedema due to a defect in brain ion and water homoeostasis. Lancet Neurology, The, 2012, 11, 973-985.	10.2	131
13	GlialCAM, a Protein Defective in a Leukodystrophy, Serves as a ClC-2 Clâ^' Channel Auxiliary Subunit. Neuron, 2012, 73, 951-961.	8.1	118
14	Investigation of LRRC8-Mediated Volume-Regulated Anion Currents in Xenopus Oocytes. Biophysical Journal, 2016, 111, 1429-1443.	0.5	94
15	Disrupting MLC1 and GlialCAM and ClC-2 interactions in leukodystrophy entails glial chloride channel dysfunction. Nature Communications, 2014, 5, 3475.	12.8	92
16	The amino acid transport system y⁺L/4F2hc is a heteromultimeric complex. FASEB Journal, 1998, 12, 1319-1329.	0.5	87
17	CLC chloride channels: correlating structure with function. Current Opinion in Structural Biology, 2002, 12, 531-539.	5.7	86
18	Localization and functional analyses of the MLC1 protein involved in megalencephalic leukoencephalopathy with subcortical cysts. Human Molecular Genetics, 2004, 13, 2581-2594.	2.9	86

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19	An Intracellular Trafficking Defect in Type I Cystinuria rBAT Mutants M467T and M467K. Journal of Biological Chemistry, 1997, 272, 9543-9549.	3.4	82
20	Molecular mechanisms of MLC1 and GLIALCAM mutations in megalencephalic leukoencephalopathy with subcortical cysts. Human Molecular Genetics, 2011, 20, 3266-3277.	2.9	80
21	Lysinuric protein intolerance: mechanisms of pathophysiology. Molecular Genetics and Metabolism, 2004, 81, 27-37.	1.1	66
22	Megalencephalic leucoencephalopathy with cysts: defect in chloride currents and cell volume regulation. Brain, 2011, 134, 3342-3354.	7.6	63
23	Molecular pathogenesis of megalencephalic leukoencephalopathy with subcortical cysts: mutations in MLC1 cause folding defects. Human Molecular Genetics, 2008, 17, 3728-3739.	2.9	60
24	Knockdown of MLC1 in primary astrocytes causes cell vacuolation: A MLC disease cell model. Neurobiology of Disease, 2011, 43, 228-238.	4.4	60
25	HepaCAM controls astrocyte self-organization and coupling. Neuron, 2021, 109, 2427-2442.e10.	8.1	52
26	Insights into MLC pathogenesis: GlialCAM is an MLC1 chaperone required for proper activation of volume-regulated anion currents. Human Molecular Genetics, 2013, 22, 4405-4416.	2.9	50
27	MLC1 is associated with the Dystrophin-Glycoprotein Complex at astrocytic endfeet. Acta Neuropathologica, 2007, 114, 403-410.	7.7	49
28	Expression patterns of MLC1 protein in the central and peripheral nervous systems. Neurobiology of Disease, 2007, 26, 532-545.	4.4	48
29	Megalencephalic leukoencephalopathy with subcortical cysts protein 1 regulates glial surface localization of GLIALCAM from fish to humans. Human Molecular Genetics, 2014, 23, 5069-5086.	2.9	34
30	GlialCAM/MLC1 modulates LRRC8/VRAC currents in an indirect manner: Implications for megalencephalic leukoencephalopathy. Neurobiology of Disease, 2018, 119, 88-99.	4.4	34
31	Depolarization causes the formation of a ternary complex between GlialCAM, MLC1 and ClC-2 in astrocytes: implications in megalencephalic leukoencephalopathy. Human Molecular Genetics, 2017, 26, 2436-2450.	2.9	33
32	Leukoencephalopathyâ€causing CLCN2 mutations are associated with impaired Cl^{â”} channel function and trafficking. Journal of Physiology, 2017, 595, 6993-7008.	2.9	33
33	GlialCAM, a CLC-2 Cl - Channel Subunit, Activates the Slow Gate of CLC Chloride Channels. Biophysical Journal, 2014, 107, 1105-1116.	0.5	32
34	Cystinuria calls for heteromultimeric amino acid transporters. Current Opinion in Cell Biology, 1998, 10, 455-461.	5.4	31
35	Deficient LRRC8A-dependent volume-regulated anion channel activity is associated with male infertility in mice. JCI Insight, 2018, 3, .	5.0	29
36	Chloride Channels in Astrocytes: Structure, Roles in Brain Homeostasis and Implications in Disease. International Journal of Molecular Sciences, 2019, 20, 1034.	4.1	28

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37	Vacuolating megalencephalic leukoencephalopathy with subcortical cysts: functional studies of novel variants inMLC1. Human Mutation, 2006, 27, 292-292.	2.5	25
38	Myotonia-related mutations in the distal C-terminus of ClC-1 and ClC-0 chloride channels affect the structure of a poly-proline helix. Biochemical Journal, 2007, 403, 79-87.	3.7	23
39	Identification and Functional Characterization ofCLCN1Mutations Found in Nondystrophic Myotonia Patients. Human Mutation, 2016, 37, 74-83.	2.5	23
40	Expanding the spectrum of megalencephalic leukoencephalopathy with subcortical cysts in two patients with GLIALCAM mutations. Neurogenetics, 2014, 15, 41-48.	1.4	22
41	Postnatal development of the astrocyte perivascular MLC1/GlialCAM complex defines a temporal window for the gliovascular unit maturation. Brain Structure and Function, 2019, 224, 1267-1278.	2.3	22
42	Drosophila ClCâ€a is required in glia of the stem cell niche for proper neurogenesis and wiring of neural circuits. Glia, 2019, 67, 2374-2398.	4.9	21
43	Structural determinants of interaction, trafficking and function in the ClCâ€2/MLC1 subunit GlialCAM involved in leukodystrophy. Journal of Physiology, 2015, 593, 4165-4180.	2.9	19
44	Megalencephalic leukoencephalopathy with subcortical cysts: A personal biochemical retrospective. European Journal of Medical Genetics, 2018, 61, 50-60.	1.3	19
45	Megalencephalic Leukoencephalopathy with Subcortical Cysts Protein-1 (MLC1) Counteracts Astrocyte Activation in Response to Inflammatory Signals. Molecular Neurobiology, 2019, 56, 8237-8254.	4.0	19
46	Megalencephalic leukoencephalopathy with subcortical cysts is a developmental disorder of the gliovascular unit. ELife, 2021, 10, .	6.0	19
47	Regulatoryâ€“auxiliary subunits of CLC chloride channelâ€“transport proteins. Journal of Physiology, 2015, 593, 4111-4127.	2.9	17
48	Identification and characterization of the zebrafish ClC-2 chloride channel orthologs. Pflugers Archiv European Journal of Physiology, 2015, 467, 1769-1781.	2.8	17
49	Cisplatin activates volume sensitive LRRC8 channel mediated currents in Xenopus oocytes. Channels, 2017, 11, 254-260.	2.8	17
50	Functional Analyses of Mutations inHEPACAMCausing Megalencephalic Leukoencephalopathy. Human Mutation, 2014, 35, 1175-1178.	2.5	16
51	Unique variants in CLCN3, encoding an endosomal anion/proton exchanger, underlie a spectrum of neurodevelopmental disorders. American Journal of Human Genetics, 2021, 108, 1450-1465.	6.2	16
52	Megalencephalic Leukoencephalopathy: Insights Into Pathophysiology and Perspectives for Therapy. Frontiers in Cellular Neuroscience, 2020, 14, 627887.	3.7	14
53	Expression of LRRC8/VRAC Currents in Xenopus Oocytes: Advantages and Caveats. International Journal of Molecular Sciences, 2018, 19, 719.	4.1	12
54	Identification of the GlialCAM interactome: the G protein-coupled receptors GPRC5B and GPR37L1 modulate megalencephalic leukoencephalopathy proteins. Human Molecular Genetics, 2021, 30, 1649-1665.	2.9	12

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55	Structural basis for the dominant or recessive character of GLIALCAM mutations found in leukodystrophies. Human Molecular Genetics, 2020, 29, 1107-1120.	2.9	10
56	Comparison of zebrafish and mice knockouts for Megalencephalic Leukoencephalopathy proteins indicates that GlialCAM/MLC1 forms a functional unit. Orphanet Journal of Rare Diseases, 2019, 14, 268.	2.7	9
57	GLIALCAM, A Glial Cell Adhesion Molecule Implicated in Neurological Disease. Advances in Neurobiology, 2014, 8, 47-59.	1.8	9
58	A modification of the split-tobacco etch virus method for monitoring interactions between membrane proteins in mammalian cells. Analytical Biochemistry, 2012, 423, 109-118.	2.4	8
59	Role of zebrafish ClCâ€K/barttin channels in apical kidney chloride reabsorption. Journal of Physiology, 2019, 597, 3969-3983.	2.9	8
60	Control of membrane protein homeostasis by a chaperone-like glial cell adhesion molecule at multiple subcellular locations. Scientific Reports, 2021, 11, 18435.	3.3	8
61	The LRRC8-mediated volume-regulated anion channel is altered in glaucoma. Scientific Reports, 2019, 9, 5392.	3.3	7
62	Cerebellar Astrocyte Transduction as Gene Therapy for Megalencephalic Leukoencephalopathy. Neurotherapeutics, 2020, 17, 2041-2053.	4.4	7
63	Ubr1-induced selective endophagy/autophagy protects against the endosomal and Ca2+-induced proteostasis disease stress. Cellular and Molecular Life Sciences, 2022, 79, 167.	5.4	6
64	CLCN1 Myotonia congenita mutation with a variable pattern of inheritance suggests a novel mechanism of dominant myotonia. Muscle and Nerve, 2018, 58, 157-160.	2.2	3
65	Split-Tobacco Etch Virus (Split-TEV) Method in G Protein-Coupled Receptor Interacting Proteins. Methods in Molecular Biology, 2021, 2268, 223-232.	0.9	3
66	Muscarinic acetylcholine receptor M1 mutations causing neurodevelopmental disorder and epilepsy. Human Mutation, 2021, 42, 1215-1220.	2.5	3
67	GPR37 Receptors and Megalencephalic Leukoencephalopathy with Subcortical Cysts. International Journal of Molecular Sciences, 2022, 23, 5528.	4.1	3
68	Novel Properties of LRRC8-Mediated VRAC Currents. Biophysical Journal, 2017, 112, 416a-417a.	0.5	1
69	Glialcam Affects CLC-Chloride Channels by Activating the Slow Gate. Biophysical Journal, 2013, 104, 628a.	0.5	0
70	Reduced Current Density and Surface Expression of a CLCN1 Mutation Causing Dominant or Recessive Myotonia in Costa Rica. Biophysical Journal, 2014, 106, 147a.	0.5	0
71	Mechanisms of Dominance of MLC2B Mutations in Glialcam, a Regulatory Subunit of the ClC-2 Chloride Channel. Biophysical Journal, 2020, 118, 266a-267a.	0.5	0
72	Dynamic expression of homeostatic ion channels in differentiated cortical astrocytes in vitro. Pflugers Archiv European Journal of Physiology, 2021, 474, 243.	2.8	0