Gustavo D Aguirre

List of Publications by Year
in descending order

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99	4,969	185998 28	102304 66
papers	citations	h-index	g-index

102	102	102	3413
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	Gene therapy restores vision in a canine model of childhood blindness. Nature Genetics, 2001, 28, 92-95.	9.4	1,130
2	Gene therapy rescues photoreceptor blindness in dogs and paves the way for treating human X-linked retinitis pigmentosa. Proceedings of the National Academy of Sciences of the United States of America, 2012, 109, 2132-2137.	3.3	237
3	Gene therapy rescues cone function in congenital achromatopsia. Human Molecular Genetics, 2010, 19, 2581-2593.	1.4	235
4	A Linkage Map of the Canine Genome. Genomics, 1997, 46, 326-336.	1.3	211
5	Different RPGR exon ORF15 mutations in Canids provide insights into photoreceptor cell degeneration. Human Molecular Genetics, 2002, 11, 993-1003.	1.4	202
6	In vivo dynamics of retinal injury and repair in the rhodopsin mutant dog model of human retinitis pigmentosa. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 5233-5238.	3.3	183
7	Canine CNGB3 mutations establish cone degeneration as orthologous to the human achromatopsia locus ACHM3. Human Molecular Genetics, 2002, 11, 1823-1833.	1.4	159
8	Naturally occurring rhodopsin mutation in the dog causes retinal dysfunction and degeneration mimicking human dominant retinitis pigmentosa. Proceedings of the National Academy of Sciences of the United States of America, 2002, 99, 6328-6333.	3.3	150
9	Breed relationships facilitate fine-mapping studies: A 7.8-kb deletion cosegregates with Collie eye anomaly across multiple dog breeds. Genome Research, 2007, 17, 1562-1571.	2.4	133
10	Progress in treating inherited retinal diseases: Early subretinal gene therapy clinical trials and candidates for future initiatives. Progress in Retinal and Eye Research, 2020, 77, 100827.	7.3	133
11	A Frameshift Mutation inRPGRExon ORF15 Causes Photoreceptor Degeneration and Inner Retina Remodeling in a Model of X-Linked Retinitis Pigmentosa. , 2006, 47, 1669.		115
12	Mutation-independent rhodopsin gene therapy by knockdown and replacement with a single AAV vector. Proceedings of the National Academy of Sciences of the United States of America, 2018, 115, E8547-E8556.	3.3	114
13	Bestrophin Gene Mutations Cause Canine Multifocal Retinopathy: A Novel Animal Model for Best Disease. , 2007, 48, 1959.		108
14	Restoration of visual function by expression of a light-gated mammalian ion channel in retinal ganglion cells or ON-bipolar cells. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, E5574-83.	3.3	104
15	Canine Retina Has a Primate Fovea-Like Bouquet of Cone Photoreceptors Which Is Affected by Inherited Macular Degenerations. PLoS ONE, 2014, 9, e90390.	1.1	100
16	Bestrophinopathy: An RPE-photoreceptor interface disease. Progress in Retinal and Eye Research, 2017, 58, 70-88.	7.3	85
17	Genetic and phenotypic variations of inherited retinal diseases in dogs: the power of within- and across-breed studies. Mammalian Genome, 2012, 23, 40-61.	1.0	78
18	Successful arrest of photoreceptor and vision loss expands the therapeutic window of retinal gene therapy to later stages of disease. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, E5844-53.	3.3	75

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19	Transient Photoreceptor Deconstruction by CNTF Enhances rAAV-Mediated Cone Functional Rescue in Late Stage CNGB3-Achromatopsia. Molecular Therapy, 2013, 21, 1131-1141.	3.7	74
20	BEST1 gene therapy corrects a diffuse retina-wide microdetachment modulated by light exposure. Proceedings of the National Academy of Sciences of the United States of America, 2018, 115, E2839-E2848.	3.3	62
21	XLPRA: A canine retinal degeneration inherited as an X-linked trait. American Journal of Medical Genetics Part A, 1994, 52, 27-33.	2.4	60
22	Optimization of Retinal Gene Therapy for X-Linked Retinitis Pigmentosa Due to RPGR Mutations. Molecular Therapy, 2017, 25, 1866-1880.	3.7	60
23	A Naturally Occurring Mutation of the Opsin Gene (T4R) in Dogs Affects Glycosylation and Stability of the G Protein-coupled Receptor. Journal of Biological Chemistry, 2004, 279, 53828-53839.	1.6	57
24	Pharmacological Modulation of Photoreceptor Outer Segment Degradation in a Human iPS Cell Model of Inherited Macular Degeneration. Molecular Therapy, 2015, 23, 1700-1711.	3.7	56
25	Long-Term Structural Outcomes of Late-Stage RPE65 Gene Therapy. Molecular Therapy, 2020, 28, 266-278.	3.7	56
26	COL9A2 and COL9A3 mutations in canine autosomal recessive oculoskeletal dysplasia. Mammalian Genome, 2010, 21, 398-408.	1.0	52
27	IQCB1 and PDE6B Mutations Cause Similar Early Onset Retinal Degenerations in Two Closely Related Terrier Dog Breeds. , 2013, 54, 7005.		40
28	Strong upregulation of inflammatory genes accompanies photoreceptor demise in canine models of retinal degeneration. PLoS ONE, 2017, 12, e0177224.	1.1	40
29	Overlap of abnormal photoreceptor development and progressive degeneration in Leber congenital amaurosis caused by NPHP5 mutation. Human Molecular Genetics, 2016, 25, 4211-4226.	1.4	35
30	scAAVengr, a transcriptome-based pipeline for quantitative ranking of engineered AAVs with single-cell resolution. ELife, 2021, 10, .	2.8	33
31	Rod and cone specific domains in the interphotoreceptor matrix. Journal of Comparative Neurology, 1991, 308, 371-380.	0.9	32
32	Application of a New Subretinal Injection Device in the Dog. Cell Transplantation, 2006, 15, 511-519.	1.2	32
33	Involvement of Innate Immune System in Late Stages of Inherited Photoreceptor Degeneration. Scientific Reports, 2017, 7, 17897.	1.6	30
34	Up-Regulation of Tumor Necrosis Factor Superfamily Genes in Early Phases of Photoreceptor Degeneration. PLoS ONE, 2013, 8, e85408.	1.1	29
35	Clinical Light Exposure, Photoreceptor Degeneration, and AP-1 Activation: A Cell Death or Cell Survival Signal in the Rhodopsin Mutant Retina?. , 2007, 48, 4907.		26
36	Assessment of visual function and retinal structure following acute light exposure in the light sensitive T4R rhodopsin mutant dog. Experimental Eye Research, 2016, 146, 341-353.	1.2	25

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37	Genetic Expression of Cyclic GMP Phosphodiesterase Activity Defines Abnormal Photoreceptor Differentiation in Neurological Mutants of Inherited Retinal Degeneration. Journal of Neurochemistry, 1986, 46, 1240-1245.	2.1	23
38	Improvement in vision: a new goal for treatment of hereditary retinal degenerations. Expert Opinion on Orphan Drugs, 2015, 3, 563-575.	0.5	23
39	Concepts and Strategies in Retinal Gene Therapy. , 2017, 58, 5399.		23
40	A hypothesis to explain the reduced blood levels of docosahexaenoic acid in inherited retinal degenerations caused by mutations in genes encoding retina-specific proteins. Lipids, 1999, 34, S235-S237.	0.7	22
41	A Naturally Occurring Canine Model of Autosomal Recessive Congenital Stationary Night Blindness. PLoS ONE, 2015, 10, e0137072.	1.1	22
42	Toxicity and Efficacy Evaluation of an Adeno-Associated Virus Vector Expressing Codon-Optimized RPGR Delivered by Subretinal Injection in a Canine Model of X-linked Retinitis Pigmentosa. Human Gene Therapy, 2020, 31, 253-267.	1.4	22
43	Canine CNGA3 Gene Mutations Provide Novel Insights into Human Achromatopsia-Associated Channelopathies and Treatment. PLoS ONE, 2015, 10, e0138943.	1.1	21
44	Safety and Efficacy of AAV5 Vectors Expressing Human or Canine CNGB3 in CNGB3-Mutant Dogs. Human Gene Therapy Clinical Development, 2017, 28, 197-207.	3.2	20
45	Gene Augmentation for X-Linked Retinitis Pigmentosa Caused by Mutations in RPGR. Cold Spring Harbor Perspectives in Medicine, 2015, 5, a017392-a017392.	2.9	19
46	Retinal histopathology of an XLRP carrier with a mutation in the RPGR exon ORF15. Experimental Eye Research, 2002, 75, 431-43.	1.2	19
47	Molecular diagnostic tests for ascertainment of genotype at the rod cone dysplasia 1 (rcd1) locus in Irish setters. Current Eye Research, 1995, 14, 243-247.	0.7	18
48	Exclusion ofRPGRIP1ins44 from Primary Causal Association with Early-Onset Coneâ€“Rod Dystrophy in Dogs. , 2012, 53, 5486.		18
49	Gene therapy reforms photoreceptor structure and restores vision in NPHP5-associated Leber congenital amaurosis. Molecular Therapy, 2021, 29, 2456-2468.	3.7	18
50	Redistribution of insoluble interphotoreceptor matrix components during photoreceptor differentiation in the mouse retina. Journal of Comparative Neurology, 1994, 345, 115-124.	0.9	17
51	Development of hereditary tapetal degeneration in the beagle dog. Current Eye Research, 1988, 7, 103-114.	0.7	16
52	Effect of diet on the fatty acid and molecular species composition of dog retina phospholipids. Lipids, 1998, 33, 1187-1193.	0.7	15
53	Genome-wide association study and whole-genome sequencing identify a deletion in LRIT3 associated with canine congenital stationary night blindness. Scientific Reports, 2019, 9, 14166.	1.6	15
54	Dose Range Finding Studies with Two RPGR Transgenes in a Canine Model of X-Linked Retinitis Pigmentosa Treated with Subretinal Gene Therapy. Human Gene Therapy, 2020, 31, 743-755.	1.4	15

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55	Exclusion of the Unfolded Protein Response in Light-Induced Retinal Degeneration in the Canine T4R RHO Model of Autosomal Dominant Retinitis Pigmentosa. PLoS ONE, 2015, 10, e0115723.	1.1	14
56	Targeting ON-bipolar cells by AAV gene therapy stably reverses LRIT3 -congenital stationary night blindness. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, e2117038119.	3.3	14
57	Variabilities in retinal function and structure in a canine model of cone-rod dystrophy associated with RPGRIP1 support multigenic etiology. Scientific Reports, 2017, 7, 12823.	1.6	13
58	Complex Structural PPT1 Variant Associated with Non-syndromic Canine Retinal Degeneration. G3: Genes, Genomes, Genetics, 2019, 9, 425-437.	0.8	13
59	An improved diagnostic test for rod cone dysplasia 1 (rcd1) using allele-specific polymerase chain reaction. Current Eye Research, 1996, 15, 583-587.	0.7	12
60	Operating in the Dark. JAMA Ophthalmology, 2008, 126, 714.	2.6	12
61	Acute and Protracted Cell Death in Light-Induced Retinal Degeneration in the Canine Model of Rhodopsin Autosomal Dominant Retinitis Pigmentosa. , 2017, 58, 270.		12
62	Photoreceptor proliferation and dysregulation of cell cycle genes in early onset inherited retinal degenerations. BMC Genomics, 2016, 17, 221.	1.2	11
63	Translational Retinal Research and Therapies. Translational Vision Science and Technology, 2018, 7, 8.	1.1	11
64	Ndr kinases regulate retinal interneuron proliferation and homeostasis. Scientific Reports, 2018, 8, 12544.	1.6	11
65	Structural Organization and Expression Pattern of the CanineRPGRIP1Isoforms in Retinal Tissue. , 2011, 52, 2989.		10
66	Rod function deficit in retained photoreceptors of patients with class B Rhodopsin mutations. Scientific Reports, 2020, 10, 12552.	1.6	10
67	Comparative localization of cystathionine beta synthases and cystathionine gamma lyase in canine, non-human primate and human retina. Experimental Eye Research, 2019, 181, 72-84.	1.2	9
68	Focal/multifocal and geographic retinal dysplasia in the dogâ€"In vivo retinal microanatomy analyses. Veterinary Ophthalmology, 2020, 23, 292-304.	0.6	9
69	Characterization of ÃŸ-glucuronidase in the retinal pigment epithelium. Current Eye Research, 1997, 16, 131-143.	0.7	8
70	Molecular studies of phenotype variation in canine RPGR-XLPRA1. Molecular Vision, 2016, 22, 319-31.	1.1	8
71	CCDC66 frameshift variant associated with a new form of early-onset progressive retinal atrophy in Portuguese Water Dogs. Scientific Reports, 2020, 10, 21162.	1.6	7
72	Candidate Genetic Modifiers for RPGR Retinal Degeneration. , 2020, 61, 20.		7

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73	Conjunctival staining with lissamine green as a predictor of tear film deficiency in dogs. Veterinary Ophthalmology, 2020, 23, 624-631.	0.6	7
74	Canine rod transducin a-1: cloning of the cDNA and evaluation of the gene as a candidate for progressive retinal atrophy. Current Eye Research, 1997, 16, 71-77.	0.7	6
75	Posterior Segment Approach for Subretinal Transplantation or Injection in the Canine Model. Cell Transplantation, 2001, 10, 317-327.	1.2	6
76	Segregation distortion in inheritance of progressive rod cone degeneration (prcd) in Miniature Poodle dogs. American Journal of Medical Genetics Part A, 1990, 35, 354-359.	2.4	5
77	Adenoviral vector-mediated ÃŸ-glucuronidase cDNA transfer to treat MPS VII RPE in vitro. Current Eye Research, 2001, 23, 357-367.	0.7	5
78	Pharmacodynamics, pharmacokinetics and biodistribution of recombinant human N-acetylgalactosamine 4-sulfatase after 6months of therapy in cats using different IV infusion durations. Molecular Genetics and Metabolism, 2016, 117, 157-163.	0.5	5
79	In-vivo longitudinal changes in thickness of the postnatal canine retina. Experimental Eye Research, 2020, 192, 107926.	1.2	5
80	Underdeveloped RPE Apical Domain Underlies Lesion Formation in Canine Bestrophinopathies. Advances in Experimental Medicine and Biology, 2018, 1074, 309-315.	0.8	5
81	Altered transsulfuration pathway enzymes and redox homeostasis in inherited retinal degenerative diseases. Experimental Eye Research, 2022, 215, 108902.	1.2	5
82	Identification of a RAPD marker linked to progressive rod-cone degeneration in dogs. Mammalian Genome, 1998, 9, 740-744.	1.0	4
83	Vitreous degeneration and associated ocular abnormalities in the dog. Veterinary Ophthalmology, 2020, 23, 219-224.	0.6	4
84	Characterization of the Canine Retinal Vasculature With Optical Coherence Tomography Angiography: Comparisons With Histology and Fluorescein Angiography. Frontiers in Neuroanatomy, 2021, 15, 785249.	0.9	4
85	Tolerability of Topical Tocilizumab Eyedrops in Dogs: A Pilot Study. Journal of Ocular Pharmacology and Therapeutics, 2017, 33, 519-524.	0.6	3
86	Short prolactin isoforms are expressed in photoreceptors of canine retinas undergoing retinal degeneration. Scientific Reports, 2021, 11, 460.	1.6	3
87	Animal Models as Tools for Screening Candidate Drugs. Retina, 2005, 25, S36-S37.	1.0	2
88	Photoreceptor Outer Segment Isolation from a Single Canine Retina for RPE Phagocytosis Assay. Advances in Experimental Medicine and Biology, 2018, 1074, 593-601.	0.8	2
89	Formal commentary. PLoS Genetics, 2020, 16, e1009059.	1.5	2
90	Is it canine DUSN?. Veterinary Ophthalmology, 2022, 25, 96-108.	0.6	2

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91	Lack of consensus on consensual response. Veterinary Ophthalmology, 2018, 21, 104-107.	0.6	1
92	Optic nerve hypoplasia in miniature poodle dogs: A preliminary genetic and candidate gene association study. Veterinary Ophthalmology, 2020, 23, 67-76.	0.6	1
93	FAM161A and TTC8 are Differentially Expressed in Non-Allelelic Early Onset Retinal Degeneration. Advances in Experimental Medicine and Biology, 2016, 854, 201-207.	0.8	1
94	Novel insights into chorioretinal and juxtapapillary colobomas by optical coherence tomography. Veterinary Ophthalmology, 2022, , .	0.6	1
95	Introducing Artur V. Cideciyan and Samuel G. Jacobson, the 2018 Recipients of the Proctor Medal. , 2019, 60, 1677.		0
96	Optic nerve colobomas associated with unilateral focal serous retinal detachment in a dog â€“ Inâ€vivo imaging and outcome following laser retinopexy. Veterinary Ophthalmology, 2021, 24, 645-652.	0.6	0
97	Retinal structural and microvascular abnormalities in retinal dysplasia imaged by OCT and OCT angiography. Veterinary Ophthalmology, 2021, , .	0.6	0
98	Monocular retinopathy of prematurityâ€like retinal vasculopathy in a dog. Veterinary Ophthalmology, 2021, , .	0.6	0
99	Remembering Alan M. Laties, MD, 1931â€“2021. , 2022, 63, 19.		0