Gustavo D Aguirre

List of Publications by Citations

97 papers	4,001 citations	29 h-index	62 g-index
102 ext. papers	4,476 ext. citations	5.4 avg, IF	4.83 L-index

#	Paper	IF	Citations
97	Gene therapy restores vision in a canine model of childhood blindness. *Nature Genetics*, **2001**, 28, 92-5	36.3	989
96	Gene therapy rescues cone function in congenital achromatopsia. *Human Molecular Genetics*, **2010**, 19, 2581-93	5.6	203
95	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, 2132-7	11.5	203
94	A linkage map of the canine genome. *Genomics*, **1997**, 46, 326-36	4.3	188
93	Different RPGR exon ORF15 mutations in Canids provide insights into photoreceptor cell degeneration. *Human Molecular Genetics*, **2002**, 11, 993-1003	5.6	159
92	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, 5233-8	11.5	153
91	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, 6328-33	11.5	138
90	Canine CNGB3 mutations establish cone degeneration as orthologous to the human achromatopsia locus ACHM3. *Human Molecular Genetics*, **2002**, 11, 1823-33	5.6	128
89	Breed relationships facilitate fine-mapping studies: a 7.8-kb deletion cosegregates with Collie eye anomaly across multiple dog breeds. *Genome Research*, **2007**, 17, 1562-71	9.7	117
88	A frameshift mutation in RPGR exon ORF15 causes photoreceptor degeneration and inner retina remodeling in a model of X-linked retinitis pigmentosa. *Investigative Ophthalmology and Visual Science*, **2006**, 47, 1669-81		99
87	Bestrophin gene mutations cause canine multifocal retinopathy: a novel animal model for best disease. *Investigative Ophthalmology and Visual Science*, **2007**, 48, 1959-67		94
86	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	11.5	80
85	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	11.5	77
84	Canine retina has a primate fovea-like bouquet of cone photoreceptors which is affected by inherited macular degenerations. *PLoS ONE*, **2014**, 9, e90390	3.7	76
83	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	20.5	76
82	Genetic and phenotypic variations of inherited retinal diseases in dogs: the power of within- and across-breed studies. *Mammalian Genome*, **2012**, 23, 40-61	3.2	66
81	Transient photoreceptor deconstruction by CNTF enhances rAAV-mediated cone functional rescue in late stage CNGB3-achromatopsia. *Molecular Therapy*, **2013**, 21, 1131-41	11.7	60

80	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	11.5	58
79	XLPRA: a canine retinal degeneration inherited as an X-linked trait. *American Journal of Medical Genetics Part A*, **1994**, 52, 27-33		58
78	Bestrophinopathy: An RPE-photoreceptor interface disease. *Progress in Retinal and Eye Research*, **2017**, 58, 70-88	20.5	57
77	Pharmacological Modulation of Photoreceptor Outer Segment Degradation in a Human iPS Cell Model of Inherited Macular Degeneration. *Molecular Therapy*, **2015**, 23, 1700-1711	11.7	50
76	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, 53828-39	5.4	48
75	COL9A2 and COL9A3 mutations in canine autosomal recessive oculoskeletal dysplasia. *Mammalian Genome*, **2010**, 21, 398-408	3.2	44
74	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	11.5	42
73	Optimization of Retinal Gene Therapy for X-Linked Retinitis Pigmentosa Due to RPGR Mutations. *Molecular Therapy*, **2017**, 25, 1866-1880	11.7	41
72	Long-Term Structural Outcomes of Late-Stage RPE65 Gene Therapy. *Molecular Therapy*, **2020**, 28, 266-278	11.7	33
71	IQCB1 and PDE6B mutations cause similar early onset retinal degenerations in two closely related terrier dog breeds **2013**, 54, 7005-19		30
70	Strong upregulation of inflammatory genes accompanies photoreceptor demise in canine models of retinal degeneration. *PLoS ONE*, **2017**, 12, e0177224	3.7	30
69	Application of a new subretinal injection device in the dog. *Cell Transplantation*, **2006**, 15, 511-9	4	29
68	Rod and cone specific domains in the interphotoreceptor matrix. *Journal of Comparative Neurology*, **1991**, 308, 371-80	3.4	27
67	Overlap of abnormal photoreceptor development and progressive degeneration in Leber congenital amaurosis caused by NPHP5 mutation. *Human Molecular Genetics*, **2016**, 25, 4211-4226	5.6	26
66	Up-regulation of tumor necrosis factor superfamily genes in early phases of photoreceptor degeneration. *PLoS ONE*, **2013**, 8, e85408	3.7	26
65	Genetic expression of cyclic GMP phosphodiesterase activity defines abnormal photoreceptor differentiation in neurological mutants of inherited retinal degeneration. *Journal of Neurochemistry* , **1986**, 46, 1240-5	6	23
64	Clinical light exposure, photoreceptor degeneration, and AP-1 activation: a cell death or cell survival signal in the rhodopsin mutant retina?. *Investigative Ophthalmology and Visual Science*, **2007**, 48, 4907-18		22
63	Involvement of Innate Immune System in Late Stages of Inherited Photoreceptor Degeneration. *Scientific Reports*, **2017**, 7, 17897	4.9	21

62	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 Suppl, S235-7	1.6	20
61	Improvement in vision: a new goal for treatment of hereditary retinal degenerations. *Expert Opinion on Orphan Drugs*, **2015**, 3, 563-575	1.1	19
60	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	3.7	19
59	Retinal histopathology of an XLRP carrier with a mutation in the RPGR exon ORF15. *Experimental Eye Research*, **2002**, 75, 431-43	3.7	18
58	Gene augmentation for X-linked retinitis pigmentosa caused by mutations in RPGR. *Cold Spring Harbor Perspectives in Medicine*, **2014**, 5, a017392	5.4	17
57	Molecular diagnostic tests for ascertainment of genotype at the rod cone dysplasia 1 (rcd1) locus in Irish setters. *Current Eye Research*, **1995**, 14, 243-7	2.9	17
56	Concepts and Strategies in Retinal Gene Therapy **2017**, 58, 5399-5411		16
55	Exclusion of RPGRIP1 ins44 from primary causal association with early-onset cone-rod dystrophy in dogs **2012**, 53, 5486-501		16
54	A Naturally Occurring Canine Model of Autosomal Recessive Congenital Stationary Night Blindness. *PLoS ONE*, **2015**, 10, e0137072	3.7	15
53	Canine CNGA3 Gene Mutations Provide Novel Insights into Human Achromatopsia-Associated Channelopathies and Treatment. *PLoS ONE*, **2015**, 10, e0138943	3.7	15
52	Effect of diet on the fatty acid and molecular species composition of dog retina phospholipids. *Lipids*, **1998**, 33, 1187-93	1.6	14
51	Redistribution of insoluble interphotoreceptor matrix components during photoreceptor differentiation in the mouse retina. *Journal of Comparative Neurology*, **1994**, 345, 115-24	3.4	14
50	Development of hereditary tapetal degeneration in the beagle dog. *Current Eye Research*, **1988**, 7, 103-14	2.9	14
49	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	13
48	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	3.7	13
47	Operating in the dark: a night-vision system for surgery in retinas susceptible to light damage. *JAMA Ophthalmology*, **2008**, 126, 714-7		12
46	An improved diagnostic test for rod cone dysplasia 1 (rcd1) using allele-specific polymerase chain reaction. *Current Eye Research*, **1996**, 15, 583-7	2.9	12
45	Photoreceptor proliferation and dysregulation of cell cycle genes in early onset inherited retinal degenerations. *BMC Genomics*, **2016**, 17, 221	4.5	10

44	Structural organization and expression pattern of the canine RPGRIP1 isoforms in retinal tissue **2011**, 52, 2989-98		10
43	Toxicity and Efficacy Evaluation of an Adeno-Associated Virus Vector Expressing Codon-Optimized Delivered by Subretinal Injection in a Canine Model of X-linked Retinitis Pigmentosa. *Human Gene Therapy*, **2020**, 31, 253-267	4.8	10
42	Complex Structural Variant Associated with Non-syndromic Canine Retinal Degeneration. *G3: Genes, Genomes, Genetics*, **2019**, 9, 425-437	3.2	10
41	Translational Retinal Research and Therapies. *Translational Vision Science and Technology*, **2018**, 7, 8	3.3	10
40	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	4.9	9
39	Characterization of beta-glucuronidase in the retinal pigment epithelium. *Current Eye Research*, **1997**, 16, 131-43	2.9	8
38	Acute and Protracted Cell Death in Light-Induced Retinal Degeneration in the Canine Model of Rhodopsin Autosomal Dominant Retinitis Pigmentosa **2017**, 58, 270-281		7
37	Molecular studies of phenotype variation in canine RPGR-XLPRA1. *Molecular Vision*, **2016**, 22, 319-31	2.3	7
36	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	3.7	7
35	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	4.9	6
34	Canine rod transducin alpha-1: cloning of the cDNA and evaluation of the gene as a candidate for progressive retinal atrophy. *Current Eye Research*, **1997**, 16, 71-7	2.9	6
33	Posterior Segment Approach for Subretinal Transplantation or Injection in the Canine Model. *Cell Transplantation*, **2001**, 10, 317-327	4	6
32	Dose Range Finding Studies with Two Transgenes in a Canine Model of X-Linked Retinitis Pigmentosa Treated with Subretinal Gene Therapy. *Human Gene Therapy*, **2020**, 31, 743-755	4.8	5
31	Adenoviral vector-mediated beta-glucuronidase cDNA transfer to treat MPS VII RPE in vitro. *Current Eye Research*, **2001**, 23, 357-67	2.9	5
30	Rod function deficit in retained photoreceptors of patients with class B Rhodopsin mutations. *Scientific Reports*, **2020**, 10, 12552	4.9	5
29	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, 157-63	3.7	4
28	Ndr kinases regulate retinal interneuron proliferation and homeostasis. *Scientific Reports*, **2018**, 8, 12544	4.9	4
27	Identification of a RAPD marker linked to progressive rod-cone degeneration in dogs. *Mammalian Genome*, **1998**, 9, 740-4	3.2	4

26	Segregation distortion in inheritance of progressive rod cone degeneration (prcd) in miniature poodle dogs. *American Journal of Medical Genetics Part A*, **1990**, 35, 354-9		4
25	Underdeveloped RPE Apical Domain Underlies Lesion Formation in Canine Bestrophinopathies. *Advances in Experimental Medicine and Biology*, **2018**, 1074, 309-315	3.6	4
24	scAAVengr, a transcriptome-based pipeline for quantitative ranking of engineered AAVs with single-cell resolution. *ELife*, **2021**, 10,	8.9	3
23	Focal/multifocal and geographic retinal dysplasia in the dog-In vivo retinal microanatomy analyses. *Veterinary Ophthalmology*, **2020**, 23, 292-304	1.4	3
22	Gene therapy reforms photoreceptor structure and restores vision in NPHP5-associated Leber congenital amaurosis. *Molecular Therapy*, **2021**, 29, 2456-2468	11.7	3
21	In-vivo longitudinal changes in thickness of the postnatal canine retina. *Experimental Eye Research*, **2020**, 192, 107926	3.7	2
20	Tolerability of Topical Tocilizumab Eyedrops in Dogs: A Pilot Study. *Journal of Ocular Pharmacology and Therapeutics*, **2017**, 33, 519-524	2.6	2
19	Formal commentary. *PLoS Genetics*, **2020**, 16, e1009059	6	2
18	Candidate Genetic Modifiers for RPGR Retinal Degeneration **2020**, 61, 20		2
17	Conjunctival staining with lissamine green as a predictor of tear film deficiency in dogs. *Veterinary Ophthalmology*, **2020**, 23, 624-631	1.4	1
16	Photoreceptor Outer Segment Isolation from a Single Canine Retina for RPE Phagocytosis Assay. *Advances in Experimental Medicine and Biology*, **2018**, 1074, 593-601	3.6	1
15	Lack of consensus on consensual response. *Veterinary Ophthalmology*, **2018**, 21, 104-107	1.4	1
14	Animal models as tools for screening candidate drugs. *Retina*, **2005**, 25, S36-S37	3.6	1
13	Altered transsulfuration pathway enzymes and redox homeostasis in inherited retinal degenerative diseases.. *Experimental Eye Research*, **2021**, 215, 108902	3.7	1
12	FAM161A and TTC8 are Differentially Expressed in Non-Allelelic Early Onset Retinal Degeneration. *Advances in Experimental Medicine and Biology*, **2016**, 854, 201-7	3.6	1
11	Vitreous degeneration and associated ocular abnormalities in the dog. *Veterinary Ophthalmology*, **2020**, 23, 219-224	1.4	1
10	Characterization of the Canine Retinal Vasculature With Optical Coherence Tomography Angiography: Comparisons With Histology and Fluorescein Angiography.. *Frontiers in Neuroanatomy*, **2021**, 15, 785249	3.6	1
9	CCDC66 frameshift variant associated with a new form of early-onset progressive retinal atrophy in Portuguese Water Dogs. *Scientific Reports*, **2020**, 10, 21162	4.9	0

8	Optic nerve hypoplasia in miniature poodle dogs: A preliminary genetic and candidate gene association study. *Veterinary Ophthalmology*, **2020**, 23, 67-76	1.4	0
7	Short prolactin isoforms are expressed in photoreceptors of canine retinas undergoing retinal degeneration. *Scientific Reports*, **2021**, 11, 460	4.9	0
6	Targeting ON-bipolar cells by AAV gene therapy stably reverses -congenital stationary night blindness.. *Proceedings of the National Academy of Sciences of the United States of America*, **2022**, 119, e2117038119	11.5	0
5	Introducing Artur V. Cideciyan and Samuel G. Jacobson, the 2018 Recipients of the Proctor Medal **2019**, 60, 1677-1679		
4	Retinal structural and microvascular abnormalities in retinal dysplasia imaged by OCT and OCT angiography. *Veterinary Ophthalmology*, **2021**,	1.4	
3	Monocular retinopathy of prematurity-like retinal vasculopathy in a dog. *Veterinary Ophthalmology*, **2021**,	1.4	
2	Optic nerve colobomas associated with unilateral focal serous retinal detachment in a dog - In-vivo imaging and outcome following laser retinopexy. *Veterinary Ophthalmology*, **2021**, 24, 645-652	1.4	
1	Remembering Alan M. Laties, MD, 1931-2021. **2022**, 63, 19		