## Fumihiko Urano

List of Publications by Year in descending order

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53794 30922 12,954 104 45 102 citations h-index g-index papers 110 110 110 17194 docs citations times ranked citing authors all docs

#	Article	IF	CITATIONS
1	IRE1 couples endoplasmic reticulum load to secretory capacity by processing the XBP-1 mRNA. Nature, 2002, 415, 92-96.	27.8	2,452
2	Autophagy Is Activated for Cell Survival after Endoplasmic ReticulumStress. Molecular and Cellular Biology, 2006, 26, 9220-9231.	2.3	1,627
3	Transcriptional and Translational Control in the Mammalian Unfolded Protein Response. Annual Review of Cell and Developmental Biology, 2002, 18, 575-599.	9.4	838
4	Measuring ER Stress and the Unfolded Protein Response Using Mammalian Tissue Culture System. Methods in Enzymology, 2011, 490, 71-92.	1.0	721
5	Thioredoxin-Interacting Protein Mediates ER Stress-Induced $\hat{l}^2$ Cell Death through Initiation of the Inflammasome. Cell Metabolism, 2012, 16, 265-273.	16.2	568
6	Compartment-specific perturbation of protein handling activates genes encoding mitochondrial chaperones. Journal of Cell Science, 2004, 117, 4055-4066.	2.0	522
7	Regulation of insulin biosynthesis in pancreatic beta cells by an endoplasmic reticulum-resident protein kinase IRE1. Cell Metabolism, 2006, 4, 245-254.	16.2	381
8	Wolfram syndrome 1 gene negatively regulates ER stress signaling in rodent and human cells. Journal of Clinical Investigation, 2010, 120, 744-755.	8.2	336
9	Endoplasmic reticulum stress and pancreatic $\hat{l}^2$ -cell death. Trends in Endocrinology and Metabolism, 2011, 22, 266-74.	7.1	310
10	WFS1 Is a Novel Component of the Unfolded Protein Response and Maintains Homeostasis of the Endoplasmic Reticulum in Pancreatic $\hat{l}^2$ -Cells. Journal of Biological Chemistry, 2005, 280, 39609-39615.	3.4	295
11	Transcriptional Regulation of VEGF-A by the Unfolded Protein Response Pathway. PLoS ONE, 2010, 5, e9575.	2.5	218
12	Wolfram Syndrome: Diagnosis, Management, and Treatment. Current Diabetes Reports, 2016, 16, 6.	4.2	198
13	A survival pathway for <i>Caenorhabditis elegans</i> with a blocked unfolded protein response. Journal of Cell Biology, 2002, 158, 639-646.	5.2	181
14	The Role of IRE1α in the Degradation of Insulin mRNA in Pancreatic β-Cells. PLoS ONE, 2008, 3, e1648.	2.5	162
15	A Novel Chimera Gene betweenEWSandE1A-F, Encoding the Adenovirus E1A Enhancer-Binding Protein, in Extraosseous Ewing's Sarcoma. Biochemical and Biophysical Research Communications, 1996, 219, 608-612.	2.1	153
16	Fluoride Induces Endoplasmic Reticulum Stress in Ameloblasts Responsible for Dental Enamel Formation. Journal of Biological Chemistry, 2005, 280, 23194-23202.	3.4	147
17	Poly-dipeptides encoded by the C9ORF72 repeats block global protein translation. Human Molecular Genetics, 2016, 25, 1803-1813.	2.9	146
18	Endoplasmic reticulum stress in $\hat{l}^2$ -cells and development of diabetes. Current Opinion in Pharmacology, 2009, 9, 763-770.	3.5	139

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19	FUS/TLS assembles into stress granules and is a prosurvival factor during hyperosmolar stress. Journal of Cellular Physiology, 2013, 228, 2222-2231.	4.1	139
20	Upregulation of BiP and CHOP by the unfolded-protein response is independent of presenilin expression. Nature Cell Biology, 2000, 2, 863-870.	10.3	136
21	Intermittent fasting preserves beta-cell mass in obesity-induced diabetes via the autophagy-lysosome pathway. Autophagy, 2017, 13, 1952-1968.	9.1	131
22	A calcium-dependent protease as a potential therapeutic target for Wolfram syndrome. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, E5292-301.	7.1	128
23	Gene-edited human stem cellâ $\in$ "derived $\hat{l}^2$ cells from a patient with monogenic diabetes reverse preexisting diabetes in mice. Science Translational Medicine, 2020, 12, .	12.4	123
24	Calcium Efflux From the Endoplasmic Reticulum Leads to β-Cell Death. Endocrinology, 2014, 155, 758-768.	2.8	122
25	AIP1 Is Critical in Transducing IRE1-mediated Endoplasmic Reticulum Stress Response. Journal of Biological Chemistry, 2008, 283, 11905-11912.	3.4	104
26	The IRE1αâ€"XBP1 pathway is essential for osteoblast differentiation through promoting transcription of <i>Osterix</i> . EMBO Reports, 2011, 12, 451-457.	4.5	103
27	Autosomal Dominant Diabetes Arising From a Wolfram Syndrome 1 Mutation. Diabetes, 2013, 62, 3943-3950.	0.6	100
28	High ER stress in $\hat{l}^2$ -cells stimulates intracellular degradation of misfolded insulin. Biochemical and Biophysical Research Communications, 2004, 324, 166-170.	2.1	86
29	The ATF6 pathway of the ER stress response contributes to enhanced viability in glioblastoma. Oncotarget, 2016, 7, 2080-2092.	1.8	86
30	Beta cells transfer vesicles containing insulin to phagocytes for presentation to T cells. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, E5496-502.	7.1	85
31	Rpl13a small nucleolar RNAs regulate systemic glucose metabolism. Journal of Clinical Investigation, 2016, 126, 4616-4625.	8.2	78
32	Dominant ER Stress–Inducing <i>WFS1</i> Mutations Underlie a Genetic Syndrome of Neonatal/Infancy-Onset Diabetes, Congenital Sensorineural Deafness, and Congenital Cataracts. Diabetes, 2017, 66, 2044-2053.	0.6	77
33	The Role of Nitric Oxide and the Unfolded Protein Response in Cytokine-Induced $\hat{I}^2$ -Cell Death. Diabetes, 2008, 57, 124-132.	0.6	76
34	Cdc42 and Rac1 are major contributors to the saturated fatty acid-stimulated JNK pathway in hepatocytes. Journal of Hepatology, 2012, 56, 192-198.	3.7	73
35	The binary switch that controls the life and death decisions of ER stressed $\hat{l}^2$ cells. Current Opinion in Cell Biology, 2011, 23, 207-215.	5.4	69
36	IRE1-Dependent Activation of AMPK in Response to Nitric Oxide. Molecular and Cellular Biology, 2011, 31, 4286-4297.	2.3	66

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37	XBP1 activates the transcription of its target genes via an ACGT core sequence under ER stress. Biochemical and Biophysical Research Communications, 2005, 331, 1146-1153.	2.1	64
38	Molecular Analysis of Ewing's Sarcoma: Another Fusion Gene, EWS-E1AF, Available for Diagnosis. Japanese Journal of Cancer Research, 1998, 89, 703-711.	1.7	63
39	Primary cilia control glucose homeostasis via islet paracrine interactions. Proceedings of the National Academy of Sciences of the United States of America, 2020, 117, 8912-8923.	7.1	63
40	Estrogens Promote Misfolded Proinsulin Degradation to Protect Insulin Production and Delay Diabetes. Cell Reports, 2018, 24, 181-196.	6.4	61
41	Endoplasmic Reticulum Stress-Induced Apoptosis and Autoimmunity in Diabetes. Current Molecular Medicine, 2006, 6, 71-77.	1.3	59
42	Stress hypERactivation in the $\hat{I}^2$ -cell. Islets, 2010, 2, 1-9.	1.8	57
43	Wolfram syndrome 1 and adenylyl cyclase 8 interact at the plasma membrane to regulate insulin production and secretion. Nature Cell Biology, 2012, 14, 1105-1112.	10.3	57
44	Endoplasmic reticulum stress in beta cells and autoimmune diabetes. Current Opinion in Immunology, 2016, 43, 60-66.	5.5	53
45	Valproate, a Mood Stabilizer, Induces WFS1 Expression and Modulates Its Interaction with ER Stress Protein GRP94. PLoS ONE, 2009, 4, e4134.	2.5	53
46	Mesencephalic Astrocyte–Derived Neurotrophic Factor as a Urine Biomarker for Endoplasmic Reticulum Stress–Related Kidney Diseases. Journal of the American Society of Nephrology: JASN, 2016, 27, 2974-2982.	6.1	49
47	ER Stress as a Trigger for $\hat{I}^2$ -Cell Dysfunction and Autoimmunity in Type 1 Diabetes. Diabetes, 2012, 61, 780-781.	0.6	48
48	Monogenic diabetes syndromes: Locus-specific databases for Alström, Wolfram, and Thiamine-responsive megaloblastic anemia. Human Mutation, 2017, 38, 764-777.	2.5	47
49	Cytoplasmic Polyadenylation Element Binding Protein Deficiency Stimulates PTEN and Stat3 mRNA Translation and Induces Hepatic Insulin Resistance. PLoS Genetics, 2012, 8, e1002457.	3.5	46
50	Neuroplastin Modulates Anti-inflammatory Effects of MANF. IScience, 2020, 23, 101810.	4.1	46
51	Insulin regulates carboxypeptidase E by modulating translation initiation scaffolding protein elF4G1 in pancreatic $\hat{l}^2$ cells. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, E2319-28.	7.1	42
52	High frequency of inactivation of the imprintedH19 gene in ?sporadic? hepatoblastoma. , 1999, 82, 490-497.		41
53	Discovery of endoplasmic reticulum calcium stabilizers to rescue ER-stressed podocytes in nephrotic syndrome. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 14154-14163.	7.1	39
54	Endoplasmic Reticulum Stress Signaling in Pancreatic $\hat{l}^2$ -Cells. Antioxidants and Redox Signaling, 2007, 9, 2335-2344.	5.4	37

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55	The binary switch between life and death of endoplasmic reticulum-stressed $\hat{l}^2$ cells. Current Opinion in Endocrinology, Diabetes and Obesity, 2010, 17, 107-112.	2.3	37
56	IRE1 prevents endoplasmic reticulum membrane permeabilization and cell death under pathological conditions. Science Signaling, 2015, 8, ra62.	3.6	36
57	Current Landscape of Treatments for Wolfram Syndrome. Trends in Pharmacological Sciences, 2019, 40, 711-714.	8.7	36
58	Nrf2/antioxidant pathway mediates $\hat{l}^2$ cell self-repair after damage by high-fat dietâ $\in$ "induced oxidative stress. JCI Insight, 2017, 2, .	5.0	36
59	KLF15 Is a Molecular Link between Endoplasmic Reticulum Stress and Insulin Resistance. PLoS ONE, 2013, 8, e77851.	2.5	35
60	Dorfin-CHIP chimeric proteins potently ubiquitylate and degrade familial ALS-related mutant SOD1 proteins and reduce their cellular toxicity. Neurobiology of Disease, 2007, 25, 331-341.	4.4	34
61	Calpain inhibitor and ibudilast rescue $\hat{l}^2$ cell functions in a cellular model of Wolfram syndrome. Proceedings of the National Academy of Sciences of the United States of America, 2020, 117, 17389-17398.	7.1	34
62	Wolfram syndrome 1 gene regulates pathways maintaining beta-cell health and survival. Laboratory Investigation, 2020, 100, 849-862.	3.7	34
63	Pathological endoplasmic reticulum stress mediated by the IRE1 pathway contributes to pre-insulitic beta cell apoptosis in a virus-induced rat model of type 1 diabetes. Diabetologia, 2013, 56, 2638-2646.	6.3	32
64	Protein kinase C signaling during T cell activation induces the endoplasmic reticulum stress response. Cell Stress and Chaperones, 2008, 13, 421-434.	2.9	29
65	The IRE1α-XBP1 Pathway Positively Regulates Parathyroid Hormone (PTH)/PTH-related Peptide Receptor Expression and Is Involved in PTH-induced Osteoclastogenesis. Journal of Biological Chemistry, 2013, 288, 1691-1695.	3.4	29
66	Neuroimaging evidence of deficient axon myelination in Wolfram syndrome. Scientific Reports, 2016, 6, 21167.	3.3	28
67	Targeting Cellular Calcium Homeostasis to Prevent Cytokine-Mediated Beta Cell Death. Scientific Reports, 2017, 7, 5611.	3.3	28
68	Wolfram Syndrome iPS Cells: The First Human Cell Model of Endoplasmic Reticulum Disease. Diabetes, 2014, 63, 844-846.	0.6	27
69	Transcriptional Regulation of X-Box-binding Protein One (XBP1) by Hepatocyte Nuclear Factor 4α (HNF4Î') Is Vital to Beta-cell Function. Journal of Biological Chemistry, 2016, 291, 6146-6157.	3.4	25
70	Novel Treatment of Chronic Graft-Versus-Host Disease in Mice Using the ER Stress Reducer 4-Phenylbutyric Acid. Scientific Reports, 2017, 7, 41939.	3.3	25
71	A phase 1b/2a clinical trial of dantrolene sodium in patients with Wolfram syndrome. JCI Insight, 2021, 6, .	5.0	24
72	Low serum osteocalcin concentration is associated with incident type 2 diabetes mellitus in Japanese women. Journal of Bone and Mineral Metabolism, 2018, 36, 470-477.	2.7	23

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73	Wolframin is a novel regulator of tau pathology and neurodegeneration. Acta Neuropathologica, 2022, 143, 547-569.	7.7	22
74	A Novel Role for the Centrosomal Protein, Pericentrin, in Regulation of Insulin Secretory Vesicle Docking in Mouse Pancreatic $\hat{l}^2$ -cells. PLoS ONE, 2010, 5, e11812.	2.5	19
75	Transmission of proteotoxicity across cellular compartments. Genes and Development, 2002, 16, 1307-1313.	5.9	18
76	Monogenic and syndromic diabetes due to endoplasmic reticulum stress. Journal of Diabetes and Its Complications, 2021, 35, 107618.	2.3	18
77	A target-agnostic screen identifies approved drugs to stabilize the endoplasmic reticulum-resident proteome. Cell Reports, 2021, 35, 109040.	6.4	18
78	Lack of matrix metalloproteinase (MMP)-1 and -3 expression in Ewing sarcoma may be due to loss of accessibility of the MMP regulatory element to the specific fusion protein in vivo. Biochemical and Biophysical Research Communications, 2002, 293, 61-71.	2.1	16
79	Establishment of a system for monitoring endoplasmic reticulum redox state in mammalian cells. Laboratory Investigation, 2013, 93, 1254-1258.	3.7	15
80	Improved function and proliferation of adult human beta cells engrafted in diabetic immunodeficient NOD-scid IL2rγnull mice treated with alogliptin. Diabetes, Metabolic Syndrome and Obesity: Targets and Therapy, 2013, 6, 493.	2.4	15
81	<i>Listeria monocytogenes</i> induces an interferonâ€enhanced activation of the integrated stress response that is detrimental for resolution of infection in mice. European Journal of Immunology, 2017, 47, 830-840.	2.9	14
82	Acute myeloid leukemia possessing jumping translocation is related to highly elevated levels of EAT/mcl-1, a Bcl-2 related gene with anti-apoptotic functions. Leukemia Research, 2000, 24, 73-77.	0.8	13
83	A case of intra-abdominal desmoplastic small-round-cell tumor with elevated serum CA125. Pediatric Surgery International, 2002, 18, 238-240.	1.4	13
84	Targeting endoplasmic reticulum to combat juvenile diabetes. Nature Reviews Endocrinology, 2014, 10, 129-130.	9.6	13
85	Loss of Function of WFS1 Causes ER Stress-Mediated Inflammation in Pancreatic Beta-Cells. Frontiers in Endocrinology, 2022, 13, 849204.	3.5	13
86	Islet cell hyperplasia in transgenic mice overexpressing EAT/mcl-1, a bcl-2 related gene. Molecular and Cellular Endocrinology, 2003, 203, 105-116.	3.2	11
87	Endoplasmic Reticulum: An Interface Between the Immune System and Metabolism. Diabetes, 2014, 63, 48-49.	0.6	11
88	A soluble endoplasmic reticulum factor as regenerative therapy for Wolfram syndrome. Laboratory Investigation, 2020, 100, 1197-1207.	3.7	9
89	Altered neuronal physiology, development, and function associated with a common chromosome 15 duplication involving CHRNA7. BMC Biology, 2021, 19, 147.	3.8	9
90	Mammalian ECD Protein Is a Novel Negative Regulator of the PERK Arm of the Unfolded Protein Response. Molecular and Cellular Biology, 2017, 37, .	2.3	7

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91	Fast and Efficient Generation of Isogenic Induced Pluripotent Stem Cell Lines Using Adenine Base Editing. CRISPR Journal, 2021, 4, 502-518.	2.9	6
92	Digenic Variants in the FGF21 Signaling Pathway Associated with Severe Insulin Resistance and Pseudoacromegaly. Journal of the Endocrine Society, 2020, 4, bvaa138.	0.2	6
93	Establishment and Characterization of a Clonal Human Extraskeletal Ewing's Sarcoma Cell Line, EES1. Tohoku Journal of Experimental Medicine, 2006, 210, 221-230.	1.2	5
94	Bisphosphonates prevent age-related weight loss in Japanese postmenopausal women. Journal of Bone and Mineral Metabolism, 2018, 36, 734-740.	2.7	4
95	Pancreatic stone protein/regenerating protein is a potential biomarker for endoplasmic reticulum stress in beta cells. Scientific Reports, 2019, 9, 5199.	3.3	3
96	Lessons from Wolfram Syndrome: Initiation of DDAVP Therapy Causes Renal Salt Wasting Due to Elevated ANP/BNP Levels, Rescued by Fludrocortisone Treatment. Indian Journal of Pediatrics, 2021, 88, 582-585.	0.8	3
97	Production of BBF2H7â€derived small peptide fragments via endoplasmic reticulum stressâ€dependent regulated intramembrane proteolysis. FASEB Journal, 2020, 34, 865-880.	0.5	2
98	Two Cases of Wolfram Syndrome Who Were Initially Diagnosed With Type 1 Diabetes. AACE Clinical Case Reports, 2022, , .	1.1	2
99	Plasma Neurofilament Light Chain Levels Are Elevated in Children and Young Adults With Wolfram Syndrome. Frontiers in Neuroscience, 2022, 16, 795317.	2.8	2
100	Research Resource: Monitoring Endoplasmic Reticulum Membrane Integrity in $\hat{I}^2$ -Cells at the Single-Cell Level. Molecular Endocrinology, 2015, 29, 473-480.	3.7	1
101	Novel elucidation and treatment of pancreatic chronic graft-versus-host disease in mice. Royal Society Open Science, 2018, 5, 181067.	2.4	1
102	A novel detrimental homozygous mutation in the WFS1 gene in two sisters from nonconsanguineous parents with untreated diabetes insipidus. Clinical Case Reports (discontinued), 2019, 7, 2355-2357.	0.5	1
103	Deficiency of WFS1 increases vulnerability to pathological tau in vitro and in vivo. Alzheimer's and Dementia, 2020, 16, e042085.	0.8	1
104	MON-078 WFS1 Related Disorder in A 4-Month Old Girl. Journal of the Endocrine Society, 2020, 4, .	0.2	0