

# David M Bedwell

## List of Publications by Year in Descending Order

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The third column is the impact factor (IF) of the journal, and the fourth column is the number of citations of the article.

65  
papers

4,847  
citations

37  
h-index

67  
g-index

67  
ext. papers

5,379  
ext. citations

8.5  
avg, IF

5.13  
L-index

#	Paper	IF	Citations
65	Analysis of patient-specific NF1 variants leads to functional insights for Ras signaling that can impact personalized medicine. <i>Human Mutation</i> , <b>2021</b> ,	4.7	2
64	A small molecule that induces translational readthrough of CFTR nonsense mutations by eRF1 depletion. <i>Nature Communications</i> , <b>2021</b> , 12, 4358	17.4	12
63	Mutation-Directed Therapeutics for Neurofibromatosis Type I. <i>Molecular Therapy - Nucleic Acids</i> , <b>2020</b> , 20, 739-753	10.7	8
62	Identification of the amino acids inserted during suppression of CFTR nonsense mutations and determination of their functional consequences. <i>Human Molecular Genetics</i> , <b>2017</b> , 26, 3116-3129	5.6	49
61	Mice with missense and nonsense NF1 mutations display divergent phenotypes compared with human neurofibromatosis type I. <i>DMM Disease Models and Mechanisms</i> , <b>2016</b> , 9, 759-67	4.1	16
60	Discovery of Clinically Approved Agents That Promote Suppression of Cystic Fibrosis Transmembrane Conductance Regulator Nonsense Mutations. <i>American Journal of Respiratory and Critical Care Medicine</i> , <b>2016</b> , 194, 1092-1103	10.2	57
59	Ataluren stimulates ribosomal selection of near-cognate tRNAs to promote nonsense suppression. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , <b>2016</b> , 113, 12508-12513	11.5	108
58	Both the autophagy and proteasomal pathways facilitate the Ubp3p-dependent depletion of a subset of translation and RNA turnover factors during nitrogen starvation in <i>Saccharomyces cerevisiae</i> . <i>Rna</i> , <b>2015</b> , 21, 898-910	5.8	17
57	Long-term nonsense suppression therapy moderates MPS I-H disease progression. <i>Molecular Genetics and Metabolism</i> , <b>2014</b> , 111, 374-381	3.7	34
56	Characterization of defects in ion transport and tissue development in cystic fibrosis transmembrane conductance regulator (CFTR)-knockout rats. <i>PLoS ONE</i> , <b>2014</b> , 9, e91253	3.7	109
55	Therapeutics based on stop codon readthrough. <i>Annual Review of Genomics and Human Genetics</i> , <b>2014</b> , 15, 371-94	9.7	174
54	Synthetic aminoglycosides efficiently suppress cystic fibrosis transmembrane conductance regulator nonsense mutations and are enhanced by ivacaftor. <i>American Journal of Respiratory Cell and Molecular Biology</i> , <b>2014</b> , 50, 805-16	5.7	110
53	Marked repression of CFTR mRNA in the transgenic Cftr(tm1kth) mouse model. <i>Journal of Cystic Fibrosis</i> , <b>2014</b> , 13, 351-2	4.1	3
52	Attenuation of nonsense-mediated mRNA decay enhances in vivo nonsense suppression. <i>PLoS ONE</i> , <b>2013</b> , 8, e60478	3.7	77
51	The designer aminoglycoside NB84 significantly reduces glycosaminoglycan accumulation associated with MPS I-H in the <i>Idua-W392X</i> mouse. <i>Molecular Genetics and Metabolism</i> , <b>2012</b> , 105, 116-25	3.7	62
50	Identification of eRF1 residues that play critical and complementary roles in stop codon recognition. <i>Rna</i> , <b>2012</b> , 18, 1210-21	5.8	27
49	Suppression of premature termination codons as a therapeutic approach. <i>Critical Reviews in Biochemistry and Molecular Biology</i> , <b>2012</b> , 47, 444-63	8.7	72

48	Suppression of nonsense mutations as a therapeutic approach to treat genetic diseases. <i>Wiley Interdisciplinary Reviews RNA</i> , <b>2011</b> , 2, 837-52	9.3	66
47	Suppression of CFTR premature termination codons and rescue of CFTR protein and function by the synthetic aminoglycoside NB54. <i>Journal of Molecular Medicine</i> , <b>2011</b> , 89, 1149-61	5.5	58
46	Characterization of an MPS I-H knock-in mouse that carries a nonsense mutation analogous to the human IDUA-W402X mutation. <i>Molecular Genetics and Metabolism</i> , <b>2010</b> , 99, 62-71	3.7	33
45	Recoding Therapies for Genetic Diseases. <i>Nucleic Acids and Molecular Biology</i> , <b>2010</b> , 123-146		6
44	Nonsense suppression activity of PTC124 (ataluren). <i>Proceedings of the National Academy of Sciences of the United States of America</i> , <b>2009</b> , 106, E64; author reply E65	11.5	34
43	Connection between stop codon reassignment and frequent use of shifty stop frameshifting. <i>Rna</i> , <b>2009</b> , 15, 889-97	5.8	13
42	Poly-L-aspartic acid enhances and prolongs gentamicin-mediated suppression of the CFTR-G542X mutation in a cystic fibrosis mouse model. <i>Journal of Biological Chemistry</i> , <b>2009</b> , 284, 6885-92	5.4	35
41	Distinct eRF3 requirements suggest alternate eRF1 conformations mediate peptide release during eukaryotic translation termination. <i>Molecular Cell</i> , <b>2008</b> , 30, 599-609	17.6	53
40	PTC124 is an orally bioavailable compound that promotes suppression of the human CFTR-G542X nonsense allele in a CF mouse model. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , <b>2008</b> , 105, 2064-9	11.5	200
39	Eukaryotic ribosomal RNA determinants of aminoglycoside resistance and their role in translational fidelity. <i>Rna</i> , <b>2008</b> , 14, 148-57	5.8	70
38	Clinical doses of amikacin provide more effective suppression of the human CFTR-G542X stop mutation than gentamicin in a transgenic CF mouse model. <i>Journal of Molecular Medicine</i> , <b>2006</b> , 84, 573-82	5.5	61
37	Eukaryotic release factor 1 phosphorylation by CK2 protein kinase is dynamic but has little effect on the efficiency of translation termination in <i>Saccharomyces cerevisiae</i> . <i>Eukaryotic Cell</i> , <b>2006</b> , 5, 1378-87		16
36	Tpa1p is part of an mRNP complex that influences translation termination, mRNA deadenylation, and mRNA turnover in <i>Saccharomyces cerevisiae</i> . <i>Molecular and Cellular Biology</i> , <b>2006</b> , 26, 5237-48	4.8	43
35	Distinct paths to stop codon reassignment by the variant-code organisms <i>Tetrahymena</i> and <i>Euplotes</i> . <i>Molecular and Cellular Biology</i> , <b>2006</b> , 26, 438-47	4.8	46
34	Aminoglycosides as potential pharmacogenetic agents in the treatment of Hailey-Hailey disease. <i>Journal of Investigative Dermatology</i> , <b>2006</b> , 126, 229-31	4.3	51
33	Discrimination between defects in elongation fidelity and termination efficiency provides mechanistic insights into translational readthrough. <i>Journal of Molecular Biology</i> , <b>2005</b> , 348, 801-15	6.5	87
32	Inhibition of phosphoglucomutase activity by lithium alters cellular calcium homeostasis and signaling in <i>Saccharomyces cerevisiae</i> . <i>American Journal of Physiology - Cell Physiology</i> , <b>2005</b> , 289, C58-67	5.4	18
31	Pharmacological Suppression of Premature Stop Mutations that Cause Genetic Diseases. <i>Current Pharmacogenomics and Personalized Medicine: the International Journal for Expert Reviews in Pharmacogenomics</i> , <b>2005</b> , 3, 259-269		22

30	GTP hydrolysis by eRF3 facilitates stop codon decoding during eukaryotic translation termination. <i>Molecular and Cellular Biology</i> , <b>2004</b> , 24, 7769-78	4.8	139
29	Leaky termination at premature stop codons antagonizes nonsense-mediated mRNA decay in <i>S. cerevisiae</i> . <i>Rna</i> , <b>2004</b> , 10, 691-703	5.8	127
28	The Ca <sup>2+</sup> homeostasis defects in a <i>pgm2Delta</i> strain of <i>Saccharomyces cerevisiae</i> are caused by excessive vacuolar Ca <sup>2+</sup> uptake mediated by the Ca <sup>2+</sup> -ATPase Pmc1p. <i>Journal of Biological Chemistry</i> , <b>2004</b> , 279, 38495-502	5.4	15
27	A <i>Saccharomyces cerevisiae</i> mutant unable to convert glucose to glucose-6-phosphate accumulates excessive glucose in the endoplasmic reticulum due to core oligosaccharide trimming. <i>Eukaryotic Cell</i> , <b>2003</b> , 2, 534-41		15
26	Extracellular Ca(2+) sensing contributes to excess Ca(2+) accumulation and vacuolar fragmentation in a <i>pmr1Delta</i> mutant of <i>S. cerevisiae</i> . <i>Journal of Cell Science</i> , <b>2003</b> , 116, 1637-46	5.3	53
25	Hexose phosphorylation and the putative calcium channel component Mid1p are required for the hexose-induced transient elevation of cytosolic calcium response in <i>Saccharomyces cerevisiae</i> . <i>Molecular Microbiology</i> , <b>2002</b> , 44, 1299-308	4.1	31
24	Clinically relevant aminoglycosides can suppress disease-associated premature stop mutations in the IDUA and P53 cDNAs in a mammalian translation system. <i>Journal of Molecular Medicine</i> , <b>2002</b> , 80, 367-76	5.5	108
23	Aminoglycoside suppression of a premature stop mutation in a <i>Cftr</i> <sup>-/-</sup> mouse carrying a human CFTR-G542X transgene. <i>Journal of Molecular Medicine</i> , <b>2002</b> , 80, 595-604	5.5	138
22	Intracellular glucose 1-phosphate and glucose 6-phosphate levels modulate Ca <sup>2+</sup> homeostasis in <i>Saccharomyces cerevisiae</i> . <i>Journal of Biological Chemistry</i> , <b>2002</b> , 277, 45751-8	5.4	30
21	Overproduction of PDR3 suppresses mitochondrial import defects associated with a TOM70 null mutation by increasing the expression of TOM72 in <i>Saccharomyces cerevisiae</i> . <i>Molecular and Cellular Biology</i> , <b>2001</b> , 21, 7576-86	4.8	15
20	Gentamicin-mediated suppression of Hurler syndrome stop mutations restores a low level of alpha-L-iduronidase activity and reduces lysosomal glycosaminoglycan accumulation. <i>Human Molecular Genetics</i> , <b>2001</b> , 10, 291-9	5.6	116
19	Evidence that systemic gentamicin suppresses premature stop mutations in patients with cystic fibrosis. <i>American Journal of Respiratory and Critical Care Medicine</i> , <b>2001</b> , 163, 1683-92	10.2	211
18	Aminoglycoside antibiotics mediate context-dependent suppression of termination codons in a mammalian translation system. <i>Rna</i> , <b>2000</b> , 6, 1044-55	5.8	272
17	Loss of the major isoform of phosphoglucomutase results in altered calcium homeostasis in <i>Saccharomyces cerevisiae</i> . <i>Journal of Biological Chemistry</i> , <b>2000</b> , 275, 5431-40	5.4	45
16	The Golgi apparatus plays a significant role in the maintenance of Ca <sup>2+</sup> homeostasis in the <i>vps33Delta</i> vacuolar biogenesis mutant of <i>Saccharomyces cerevisiae</i> . <i>Journal of Biological Chemistry</i> , <b>1999</b> , 274, 5939-47	5.4	54
15	Mutations in the yeast Hsp40 chaperone protein Ydj1 cause defects in Axl1 biogenesis and pro-a-factor processing. <i>Journal of Biological Chemistry</i> , <b>1999</b> , 274, 34396-402	5.4	20
14	The vacuolar Ca <sup>2+</sup> /H <sup>+</sup> exchanger Vcx1p/Hum1p tightly controls cytosolic Ca <sup>2+</sup> levels in <i>S. cerevisiae</i> . <i>FEBS Letters</i> , <b>1999</b> , 451, 132-6	3.8	118
13	The amino terminus of the F1-ATPase beta-subunit precursor functions as an intramolecular chaperone to facilitate mitochondrial protein import. <i>Molecular and Cellular Biology</i> , <b>1997</b> , 17, 7169-77	4.8	19

12	Suppression of a CFTR premature stop mutation in a bronchial epithelial cell line. <i>Nature Medicine</i> , <b>1997</b> , 3, 1280-4	50.5	281
11	Mutations within the first LSGGQ motif of Ste6p cause defects in a-factor transport and mating in <i>Saccharomyces cerevisiae</i> . <i>Journal of Bacteriology</i> , <b>1996</b> , 178, 1712-9	3.5	27
10	Aminoglycoside antibiotics restore CFTR function by overcoming premature stop mutations. <i>Nature Medicine</i> , <b>1996</b> , 2, 467-9	50.5	399
9	Reply to Nonstop treatment of cystic fibrosis. <i>Nature Medicine</i> , <b>1996</b> , 2, 608-609	50.5	3
8	The posttranslational modification of phosphoglucomutase is regulated by galactose induction and glucose repression in <i>Saccharomyces cerevisiae</i> . <i>Journal of Bacteriology</i> , <b>1995</b> , 177, 3087-94	3.5	26
7	The efficiency of translation termination is determined by a synergistic interplay between upstream and downstream sequences in <i>Saccharomyces cerevisiae</i> . <i>Journal of Molecular Biology</i> , <b>1995</b> , 251, 334-45	6.5	244
6	Regulation of alpha operon gene expression in <i>Escherichia coli</i> . A novel form of translational coupling. <i>Journal of Molecular Biology</i> , <b>1987</b> , 196, 333-45	6.5	55
5	Feedback regulation of RNA polymerase subunit synthesis after the conditional overproduction of RNA polymerase in <i>Escherichia coli</i> . <i>Molecular Genetics and Genomics</i> , <b>1986</b> , 204, 17-23		37
4	Nucleotide sequence of the alpha ribosomal protein operon of <i>Escherichia coli</i> . <i>Nucleic Acids Research</i> , <b>1985</b> , 13, 3891-903	20.1	97
3	Increased expression of ribosomal genes during inhibition of ribosome assembly in <i>Escherichia coli</i> . <i>Journal of Molecular Biology</i> , <b>1985</b> , 184, 23-30	6.5	26
2	The spc ribosomal protein operon of <i>Escherichia coli</i> : sequence and cotranscription of the ribosomal protein genes and a protein export gene. <i>Nucleic Acids Research</i> , <b>1983</b> , 11, 2599-616	20.1	262
1	Characterization of a HindIII-generated DNA fragment carrying the glutamine synthetase gene of <i>Salmonella typhimurium</i> . <i>Gene</i> , <b>1980</b> , 11, 227-37	3.8	15