

# William G Kaelin

## List of Publications by Year in descending order

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112  
papers

25,837  
citations

17405

63  
h-index

26548

107  
g-index

155  
all docs

155  
docs citations

155  
times ranked

31668  
citing authors

| #  | ARTICLE  | IF   | CITATIONS |
|----|--|------|-----------|
| 1  | Oxygen Sensing by Metazoans: The Central Role of the HIF Hydroxylase Pathway. <i>Molecular Cell</i> , 2008, 30, 393-402.   | 4.5  | 2,614     |
| 2  | Ubiquitination of hypoxia-inducible factor requires direct binding to the $\beta^2$ -domain of the von Hippel-Lindau protein. <i>Nature Cell Biology</i> , 2000, 2, 423-427. | 4.6  | 1,423     |
| 3  | The Concept of Synthetic Lethality in the Context of Anticancer Therapy. <i>Nature Reviews Cancer</i> , 2005, 5, 689-698.  | 12.8 | 1,278     |
| 4  | The Myeloma Drug Lenalidomide Promotes the Cereblon-Dependent Destruction of Ikaros Proteins. <i>Science</i> , 2014, 343, 305-309.   | 6.0  | 1,196     |
| 5  | Genomic correlates of response to immune checkpoint therapies in clear cell renal cell carcinoma. <i>Science</i> , 2018, 359, 801-806.                                       | 6.0  | 898       |
| 6  | The tyrosine kinase c-Abl regulates p73 in apoptotic response to cisplatin-induced DNA damage. <i>Nature</i> , 1999, 399, 806-809.   | 13.7 | 863       |
| 7  | Structure of the VHL-ElonginC-ElonginB Complex: Implications for VHL Tumor Suppressor Function. <i>Science</i> , 1999, 284, 455-461.   | 6.0  | 793       |
| 8  | Molecular basis of the VHL hereditary cancer syndrome. <i>Nature Reviews Cancer</i> , 2002, 2, 673-682.  | 12.8 | 767       |
| 9  | Influence of Metabolism on Epigenetics and Disease. <i>Cell</i> , 2013, 153, 56-69.  | 13.5 | 729       |
| 10 | Inhibition of HIF is necessary for tumor suppression by the von Hippel-Lindau protein. <i>Cancer Cell</i> , 2002, 1, 237-246.  | 7.7  | 695       |
| 11 | Structure of an HIF-1 $\alpha$ -pVHL Complex: Hydroxyproline Recognition in Signaling. <i>Science</i> , 2002, 296, 1886-1889.  | 6.0  | 679       |
| 12 | Tumour suppression by the human von Hippel-Lindau gene product. <i>Nature Medicine</i> , 1995, 1, 822-826.   | 15.2 | 636       |
| 13 | Transformation by the (R)-enantiomer of 2-hydroxyglutarate linked to EGLN activation. <i>Nature</i> , 2012, 483, 484-488.  | 13.7 | 630       |
| 14 | (R)-2-Hydroxyglutarate Is Sufficient to Promote Leukemogenesis and Its Effects Are Reversible. <i>Science</i> , 2013, 339, 1621-1625.  | 6.0  | 624       |
| 15 | The von Hippel-Lindau tumour suppressor protein: O <sub>2</sub> sensing and cancer. <i>Nature Reviews Cancer</i> , 2008, 8, 865-873.   | 12.8 | 616       |
| 16 | Inhibition of HIF2 $\beta$ Is Sufficient to Suppress pVHL-Defective Tumor Growth. <i>PLoS Biology</i> , 2003, 1, e83.  | 2.6  | 516       |
| 17 | Neuronal apoptosis linked to EglN3 prolyl hydroxylase and familial pheochromocytoma genes: Developmental culling and cancer. <i>Cancer Cell</i> , 2005, 8, 155-167.          | 7.7  | 494       |
| 18 | What a difference a hydroxyl makes: mutant IDH, (R)-2-hydroxyglutarate, and cancer. <i>Genes and Development</i> , 2013, 27, 836-852.  | 2.7  | 491       |

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|----|---|------|-----------|
| 19 | PROLINE HYDROXYLATION AND GENE EXPRESSION. Annual Review of Biochemistry, 2005, 74, 115-128.  | 5.0  | 410       |
| 20 | The Retinoblastoma Binding Protein RBP2 Is an H3K4 Demethylase. Cell, 2007, 128, 889-900.   | 13.5 | 399       |
| 21 | Genetic and Functional Studies Implicate <i>HIF1<math>\alpha</math></i> as a 14q Kidney Cancer Suppressor Gene. Cancer Discovery, 2011, 1, 222-235.   | 7.7  | 347       |
| 22 | On-target efficacy of a HIF-2 $\alpha$ antagonist in preclinical kidney cancer models. Nature, 2016, 539, 107-111.  | 13.7 | 341       |
| 23 | A genetic mechanism for Tibetan high-altitude adaptation. Nature Genetics, 2014, 46, 951-956.   | 9.4  | 322       |
| 24 | A common E2F-1 and p73 pathway mediates cell death induced by TCR activation. Nature, 2000, 407, 642-645.   | 13.7 | 309       |
| 25 | von Hippel-Lindau Disease. Annual Review of Pathology: Mechanisms of Disease, 2007, 2, 145-173.   | 9.6  | 293       |
| 26 | Histone demethylase KDM6A directly senses oxygen to control chromatin and cell fate. Science, 2019, 363, 1217-1222.   | 6.0  | 281       |
| 27 | The von Hippel-Lindau Tumor Suppressor Protein and Clear Cell Renal Carcinoma: Fig. 1.. Clinical Cancer Research, 2007, 13, 680s-684s.  | 3.2  | 275       |
| 28 | Mouse model for noninvasive imaging of HIF prolyl hydroxylase activity: Assessment of an oral agent that stimulates erythropoietin production. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 105-110. | 3.3  | 274       |
| 29 | Genomic sequencing of colorectal adenocarcinomas identifies a recurrent VTI1A-TCF7L2 fusion. Nature Genetics, 2011, 43, 964-968.  | 9.4  | 270       |
| 30 | The Von Hippel-Lindau Tumor Suppressor Gene and Kidney Cancer: Fig. 1.. Clinical Cancer Research, 2004, 10, 6290S-6295S.  | 3.2  | 268       |
| 31 | Targeting the HIF-2 $\alpha$ -VEGF axis in renal cell carcinoma. Nature Medicine, 2020, 26, 1519-1530.  | 15.2 | 248       |
| 32 | Fumarate and Succinate Regulate Expression of Hypoxia-inducible Genes via TET Enzymes. Journal of Biological Chemistry, 2016, 291, 4256-4265.   | 1.6  | 234       |
| 33 | Transaminase Inhibition by 2-Hydroxyglutarate Impairs Glutamate Biosynthesis and Redox Homeostasis in Glioma. Cell, 2018, 175, 101-116.e25.   | 13.5 | 234       |
| 34 | Binding of pRB to the PHD Protein RBP2 Promotes Cellular Differentiation. Molecular Cell, 2005, 18, 623-635.  | 4.5  | 215       |
| 35 | Failure to prolyl hydroxylate hypoxia-inducible factor $\alpha$ phenocopies VHL inactivation in vivo. EMBO Journal, 2006, 25, 4650-4662.  | 3.5  | 210       |
| 36 | The von Hippel-Lindau protein, HIF hydroxylation, and oxygen sensing. Biochemical and Biophysical Research Communications, 2005, 338, 627-638.  | 1.0  | 197       |

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|----|---|------|-----------|
| 37 | The EGLN-HIF O <sub>2</sub> -Sensing System: Multiple Inputs and Feedbacks. <i>Molecular Cell</i> , 2017, 66, 772-779.  | 4.5  | 192       |
| 38 | Paracrine Induction of HIF by Glutamate in Breast Cancer: EglN1 Senses Cysteine. <i>Cell</i> , 2016, 166, 126-139.  | 13.5 | 187       |
| 39 | The p53 gene family. <i>Oncogene</i> , 1999, 18, 7701-7705.   | 2.6  | 175       |
| 40 | pVHL suppresses kinase activity of Akt in a proline-hydroxylation-dependent manner. <i>Science</i> , 2016, 353, 929-932.  | 6.0  | 165       |
| 41 | pVHL Acts as an Adaptor to Promote the Inhibitory Phosphorylation of the NF- $\kappa$ B Agonist Card9 by CK2. <i>Molecular Cell</i> , 2007, 28, 15-27.  | 4.5  | 163       |
| 42 | Tumor-selective transgene expression in vivo mediated by an E2F-responsive adenoviral vector. <i>Nature Medicine</i> , 1997, 3, 1145-1149.  | 15.2 | 158       |
| 43 | Use and Abuse of RNAi to Study Mammalian Gene Function. <i>Science</i> , 2012, 337, 421-422.  | 6.0  | 158       |
| 44 | ROS: Really involved in Oxygen Sensing. <i>Cell Metabolism</i> , 2005, 1, 357-358.  | 7.2  | 150       |
| 45 | Loss of the retinoblastoma binding protein 2 (RBP2) histone demethylase suppresses tumorigenesis in mice lacking <i>Rb1</i> or <i>Men1</i> . <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2011, 108, 13379-13386. | 3.3  | 143       |
| 46 | How oxygen makes its presence felt. <i>Genes and Development</i> , 2002, 16, 1441-1445.   | 2.7  | 138       |
| 47 | CDK7 Inhibition Potentiates Genome Instability Triggering Anti-tumor Immunity in Small Cell Lung Cancer. <i>Cancer Cell</i> , 2020, 37, 37-54.e9.   | 7.7  | 138       |
| 48 | Kinase requirements in human cells: III. Altered kinase requirements in <i>VHL</i> cancer cells detected in a pilot synthetic lethal screen. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2008, 105, 16484-16489. | 3.3  | 137       |
| 49 | SQSTM1 Is a Pathogenic Target of 5q Copy Number Gains in Kidney Cancer. <i>Cancer Cell</i> , 2013, 24, 738-750.   | 7.7  | 135       |
| 50 | Common pitfalls in preclinical cancer target validation. <i>Nature Reviews Cancer</i> , 2017, 17, 441-450.  | 12.8 | 134       |
| 51 | VHL substrate transcription factor ZHX2 as an oncogenic driver in clear cell renal cell carcinoma. <i>Science</i> , 2018, 361, 290-295.   | 6.0  | 134       |
| 52 | Inactivation of the PBRM1 tumor suppressor gene amplifies the HIF-response in VHL clear cell renal carcinoma. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2017, 114, 1027-1032.                                  | 3.3  | 126       |
| 53 | Deubiquitinases Maintain Protein Homeostasis and Survival of Cancer Cells upon Glutathione Depletion. <i>Cell Metabolism</i> , 2019, 29, 1166-1181.e6.  | 7.2  | 121       |
| 54 | Control of Cyclin D1 and Breast Tumorigenesis by the EglN2 Prolyl Hydroxylase. <i>Cancer Cell</i> , 2009, 16, 413-424.  | 7.7  | 120       |

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|----|---|------|-----------|
| 55 | PHD3 Loss in Cancer Enables Metabolic Reliance on Fatty Acid Oxidation via Deactivation of ACC2. <i>Molecular Cell</i> , 2016, 63, 1006-1020.   | 4.5  | 120       |
| 56 | Cells Lacking the <i>RB1</i> Tumor Suppressor Gene Are Hyperdependent on Aurora B Kinase for Survival. <i>Cancer Discovery</i> , 2019, 9, 230-247.  | 7.7  | 119       |
| 57 | 2-Oxoglutarate-dependent dioxygenases in cancer. <i>Nature Reviews Cancer</i> , 2020, 20, 710-726.  | 12.8 | 119       |
| 58 | The von Hippel-Lindau Gene, Kidney Cancer, and Oxygen Sensing. <i>Journal of the American Society of Nephrology: JASN</i> , 2003, 14, 2703-2711.  | 3.0  | 115       |
| 59 | Prolyl hydroxylation by EglN2 destabilizes FOXO3a by blocking its interaction with the USP9x deubiquitinase. <i>Genes and Development</i> , 2014, 28, 1429-1444.  | 2.7  | 111       |
| 60 | EGLN1 Inhibition and Rerouting of $\alpha$ -Ketoglutarate Suffice for Remote Ischemic Protection. <i>Cell</i> , 2016, 164, 884-895.   | 13.5 | 108       |
| 61 | Treatment of kidney cancer. <i>Cancer</i> , 2009, 115, 2262-2272.   | 2.0  | 105       |
| 62 | Hypoxia-Inducible Factor Linked to Differential Kidney Cancer Risk Seen with Type 2A and Type 2B VHL Mutations. <i>Molecular and Cellular Biology</i> , 2007, 27, 5381-5392.  | 1.1  | 102       |
| 63 | Synthetic lethality: a framework for the development of wiser cancer therapeutics. <i>Genome Medicine</i> , 2009, 1, 99.  | 3.6  | 77        |
| 64 | Phosphorylation of ETS1 by Src Family Kinases Prevents Its Recognition by the COP1 Tumor Suppressor. <i>Cancer Cell</i> , 2014, 26, 222-234.  | 7.7  | 71        |
| 65 | The VHL Tumor Suppressor Gene: Insights into Oxygen Sensing and Cancer. <i>Transactions of the American Clinical and Climatological Association</i> , 2017, 128, 298-307.   | 0.9  | 70        |
| 66 | The KDM5A/RBP2 histone demethylase represses NOTCH signaling to sustain neuroendocrine differentiation and promote small cell lung cancer tumorigenesis. <i>Genes and Development</i> , 2019, 33, 1718-1738.                          | 2.7  | 65        |
| 67 | SDH5 Mutations and Familial Paraganglioma: Somewhere Warburg is Smiling. <i>Cancer Cell</i> , 2009, 16, 180-182.  | 7.7  | 58        |
| 68 | EglN2 associates with the <i>NRF1</i> complex and controls mitochondrial function in breast cancer. <i>EMBO Journal</i> , 2015, 34, 2953-2970.  | 3.5  | 58        |
| 69 | Skp2 dictates cell cycle-dependent metabolic oscillation between glycolysis and TCA cycle. <i>Cell Research</i> , 2021, 31, 80-93.  | 5.7  | 51        |
| 70 | Many vessels, faulty gene. <i>Nature</i> , 1999, 399, 203-204.  | 13.7 | 47        |
| 71 | HIF-independent synthetic lethality between CDK4/6 inhibition and VHL loss across species. <i>Science Signaling</i> , 2019, 12, .   | 1.6  | 47        |
| 72 | Inhibition of the oxygen sensor PHD2 in the liver improves survival in lactic acidosis by activating the Cori cycle. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2015, 112, 11642-11647. | 3.3  | 46        |

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|----|--|------|-----------|
| 73 | Targeting HIF2 in Clear Cell Renal Cell Carcinoma. Cold Spring Harbor Symposia on Quantitative Biology, 2016, 81, 113-121.   | 2.0  | 43        |
| 74 | The von Hippel-Lindau Tumor Suppressor Protein: An Update. Methods in Enzymology, 2007, 435, 371-383.  | 0.4  | 42        |
| 75 | Peptidic degron for IMiD-induced degradation of heterologous proteins. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 2539-2544.  | 3.3  | 41        |
| 76 | Mouse Reporter Strain for Noninvasive Bioluminescent Imaging of Cells that have Undergone Cre-Mediated Recombination. Molecular Imaging, 2003, 2, 153535002003031.   | 0.7  | 36        |
| 77 | Pathways for Oxygen Regulation and Homeostasis. JAMA - Journal of the American Medical Association, 2016, 316, 1252.   | 3.8  | 36        |
| 78 | HIF activation causes synthetic lethality between the <i>VHL</i> tumor suppressor and the <i>EZH1</i> histone methyltransferase. Science Translational Medicine, 2017, 9, .  | 5.8  | 36        |
| 79 | Belzutifan, a Potent HIF2 $\alpha$ Inhibitor, in the Pacak-Zhuang Syndrome. New England Journal of Medicine, 2021, 385, 2059-2065.   | 13.9 | 36        |
| 80 | Genetic Evidence of a Precisely Tuned Dysregulation in the Hypoxia Signaling Pathway during Oncogenesis. Cancer Research, 2014, 74, 6554-6564.   | 0.4  | 32        |
| 81 | Kidney Cancer: Now Available in a New Flavor. Cancer Cell, 2008, 14, 423-424.  | 7.7  | 31        |
| 82 | Targeting oncoproteins with a positive selection assay for protein degraders. Science Advances, 2021, 7, .   | 4.7  | 26        |
| 83 | Peptidic degron in EID1 is recognized by an SCF E3 ligase complex containing the orphan F-box protein FBXO21. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, 15372-15377.                   | 3.3  | 24        |
| 84 | Cyclin D1 suppresses retinoblastoma protein-mediated inhibition of TAFII250 kinase activity. Oncogene, 2000, 19, 5703-5711.  | 2.6  | 21        |
| 85 | Mutant p53 induces a hypoxia transcriptional program in gastric and esophageal adenocarcinoma. JCI Insight, 2019, 4, .   | 2.3  | 21        |
| 86 | BRCA1-IRIS promotes human tumor progression through PTEN blockade and HIF-1 $\alpha$ activation. Proceedings of the National Academy of Sciences of the United States of America, 2018, 115, E9600-E9609.                                | 3.3  | 20        |
| 87 | Gleevec: Prototype or Outlier?. Science Signaling, 2004, 2004, pe12-pe12.  | 1.6  | 19        |
| 88 | HIF2 Inhibitor Joins the Kidney Cancer Armamentarium. Journal of Clinical Oncology, 2018, 36, 908-910.   | 0.8  | 14        |
| 89 | The von Hippel-Lindau Tumor Suppressor Protein. Annual Review of Cancer Biology, 2018, 2, 91-109.  | 2.3  | 13        |
| 90 | Egln3 hydroxylase stabilizes BIM-EL linking VHL type 2C mutations to pheochromocytoma pathogenesis and chemotherapy resistance. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 16997-17006. | 3.3  | 13        |

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|-----|--|------|-----------|
| 91  | From Basic Science to Clinical Translation in Kidney Cancer: A Report from the Second Kidney Cancer Research Summit. <i>Clinical Cancer Research</i> , 2022, 28, 831-839.  | 3.2  | 12        |
| 92  | New cancer targets emerging from studies of the Von Hippel-Lindau tumor suppressor protein. <i>Annals of the New York Academy of Sciences</i> , 2010, 1210, 1-7.   | 1.8  | 11        |
| 93  | Sensitivity of VHL mutant kidney cancers to HIF2 inhibitors does not require an intact p53 pathway. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2022, 119, e2120403119.   | 3.3  | 11        |
| 94  | Autochthonous tumors driven by Rb1 loss have an ongoing requirement for the RBP2 histone demethylase. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2018, 115, E3741-E3748. | 3.3  | 10        |
| 95  | <i>Molecular Biology of Kidney Cancer</i> . , 2015, , 31-57.   |      | 10        |
| 96  | Analysis of von Hippel-Lindau Hereditary Cancer Syndrome: Implications of Oxygen Sensing. <i>Methods in Enzymology</i> , 2004, 381, 320-335.   | 0.4  | 9         |
| 97  | A Mesenchymal Tumor Cell State Confers Increased Dependency on the BCL-XL Antiapoptotic Protein in Kidney Cancer. <i>Clinical Cancer Research</i> , 2022, 28, 4689-4701.   | 3.2  | 5         |
| 98  | Leveraging insights into cancer metabolism—a symposium report. <i>Annals of the New York Academy of Sciences</i> , 2020, 1462, 5-13.   | 1.8  | 3         |
| 99  | Disabling Kidney Cancers Caused by Fumarate Hydratase Mutations. <i>Cancer Cell</i> , 2014, 26, 779-780.   | 7.7  | 2         |
| 100 | Climate Change. <i>JAMA - Journal of the American Medical Association</i> , 2017, 318, 611.  | 3.8  | 2         |
| 101 | DDRE-29. DE NOVO PYRIMIDINE SYNTHESIS IS A TARGETABLE VULNERABILITY IN IDH-MUTANT GLIOMA. <i>Neuro-Oncology Advances</i> , 2021, 3, i12-i13.   | 0.4  | 1         |
| 102 | Liver Specific Delivery of siRNA Targeting EGLN Prolyl Hydroxylases Activates Hepatic Erythropoietin Production and Stimulates Erythropoiesis. <i>Blood</i> , 2011, 118, 3161-3161.                                    | 0.6  | 1         |
| 103 | Enantiomer-Specific Transformation by 2HG Is Linked to Opposing Effects on $\alpha$ -Ketoglutarate-Dependent Dioxygenases. <i>Blood</i> , 2011, 118, LBA-4-LBA-4.  | 0.6  | 1         |
| 104 | A Comprehensive Study of the VHL-R200W Chuvash Polycythemia Mutation Reveals a Gradual Dysregulation of the Hypoxia Pathway in Oncogenesis. <i>Blood</i> , 2014, 124, 4020-4020.                                       | 0.6  | 1         |
| 105 | IDH Mutations, 2-Oxoglutarate-dependent Dioxygenases, and Leukemia. <i>Blood</i> , 2014, 124, SCI-6-SCI-6.   | 0.6  | 1         |
| 106 | Mutation Selective IDH Inhibitors Mediate Histone and DNA Methylation Changes. <i>Blood</i> , 2012, 120, 3509-3509.  | 0.6  | 1         |
| 107 | David M. Livingston (1941–2021). <i>Cell</i> , 2021, 184, 6007-6009.   | 13.5 | 1         |
| 108 | Senator McCain and Our Shared Humanity. <i>American Journal of Medicine</i> , 2018, 131, 216-217.  | 0.6  | 0         |

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| 109 | Enantiomer-Specific Transformation by 2HG Is Linked to Opposing Effects on $\hat{\pm}$ -Ketoglutarate-Dependent Dioxygenases. Blood, 2011, 118, LBA-4-LBA-4. | 0.6 | 0         |
| 110 | Transformation by Mutant IDH and (R)-2HG Is Reversible.. Blood, 2012, 120, 2413-2413.  | 0.6 | 0         |
| 111 | Disruption of the Ikaros-Mediated Gene Expression Program in Multiple Myeloma with Immunomodulatory Agents. Blood, 2014, 124, 420-420.                       | 0.6 | 0         |
| 112 | Targeting Oncoproteins with a Positive Selection Assay for Protein Degradors. Blood, 2020, 136, 13-14.   | 0.6 | 0         |