Simon R Johnson

List of Publications by Year
in descending order

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Version: 2024-02-01

102 papers	6,742 citations	109264 35 h-index	62565 80 g-index
104 all docs	104 docs citations	104 times ranked	5745 citing authors

#	Article	IF	Citations
1	Tuberous Sclerosis Complex Diagnostic Criteria Update: Recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference. Pediatric Neurology, 2013, 49, 243-254.	1.0	1,185
2	Tuberous Sclerosis Complex Surveillance and Management: Recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference. Pediatric Neurology, 2013, 49, 255-265.	1.0	693
3	European Respiratory Society guidelines for the diagnosis and management of lymphangioleiomyomatosis. European Respiratory Journal, 2010, 35, 14-26.	3.1	468
4	Sirolimus Therapy for Angiomyolipoma in Tuberous Sclerosis and Sporadic Lymphangioleiomyomatosis: A Phase 2 Trial. Clinical Cancer Research, 2011, 17, 4071-4081.	3.2	278
5	Official American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines: Lymphangioleiomyomatosis Diagnosis and Management. American Journal of Respiratory and Critical Care Medicine, 2016, 194, 748-761.	2.5	236
6	Updated International Tuberous Sclerosis Complex Diagnostic Criteria and Surveillance and Management Recommendations. Pediatric Neurology, 2021, 123, 50-66.	1.0	230
7	Clinical experience of lymphangioleiomyomatosis in the UK. Thorax, 2000, 55, 1052-1057.	2.7	215
8	Lymphangioleiomyomatosis. European Respiratory Journal, 2006, 27, 1056-1065.	3.1	211
9	Sirolimus Therapy in Tuberous Sclerosis or Sporadic Lymphangioleiomyomatosis. New England Journal of Medicine, 2008, 358, 200-203.	13.9	208
10	Lymphangioleiomyomatosis Diagnosis and Management: High-Resolution Chest Computed Tomography, Transbronchial Lung Biopsy, and Pleural Disease Management. An Official American Thoracic Society/Japanese Respiratory Society Clinical Practice Guideline. American Journal of Respiratory and Critical Care Medicine, 2017, 196, 1337-1348.	2.5	159
11	Survival and disease progression in UK patients with lymphangioleiomyomatosis. Thorax, 2004, 59, 800-803.	2.7	137
12	Effects of Growth Factors and Extracellular Matrix on Survival of Human Airway Smooth Muscle Cells. American Journal of Respiratory Cell and Molecular Biology, 2001, 25, 569-576.	1.4	134
13	Use of variability in national and regional data to estimate the prevalence of lymphangioleiomyomatosis. QJM - Monthly Journal of the Association of Physicians, 2011, 104, 971-979.	0.2	121
14	Extracellular Matrix Cross-Linking Enhances Fibroblast Growth and Protects against Matrix Proteolysis in Lung Fibrosis. American Journal of Respiratory Cell and Molecular Biology, 2018, 58, 594-603.	1.4	111
15	Presence of a prothrombotic state in people with idiopathic pulmonary fibrosis: a population-based caseâ€“control study. Thorax, 2014, 69, 207-215.	2.7	106
16	Synthetic functions of airway smooth muscle in asthma. Trends in Pharmacological Sciences, 1997, 18, 288-292.	4.0	98
17	Synthetic functions of airway smooth muscle in asthma. Trends in Pharmacological Sciences, 1997, 18, 288-292.	4.0	95
18	Lymphangioleiomyomatosis. Clinics in Chest Medicine, 2016, 37, 389-403.	0.8	94

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19	Accelerated extracellular matrix turnover during exacerbations of COPD. Respiratory Research, 2015, 16, 69.	1.4	78
20	Emerging clinical picture of lymphangioleiomyomatosis. Thorax, 2005, 60, 875-879.	2.7	77
21	Pregnancy experiences among women with lymphangioleiomyomatosis. Respiratory Medicine, 2009, 103, 766-772.	1.3	70
22	Association of MMP - 12 polymorphisms with severe and very severe COPD: A case control study of MMPs - 1, 9 and 12in a European population. BMC Medical Genetics, 2010, 11, 7.	2.1	70
23	Lymphangioleiomyomatosis: pathogenesis, clinical features, diagnosis, and management. Lancet Respiratory Medicine,the, 2021, 9, 1313-1327.	5.2	67
24	Lung function response and side effects to rapamycin for lymphangioleiomyomatosis: a prospective national cohort study. Thorax, 2018, 73, 369-375.	2.7	65
25	Matrix metalloproteinase expression and activity in human airway smooth muscle cells. British Journal of Pharmacology, 2004, 142, 1318-1324.	2.7	62
26	A 2-year randomised placebo-controlled trial of doxycycline for lymphangioleiomyomatosis. European Respiratory Journal, 2014, 43, 1114-1123.	3.1	60
27	Autocrine production of matrix metalloproteinase-2 is required for human airway smooth muscle proliferation. American Journal of Physiology - Lung Cellular and Molecular Physiology, 1999, 277, L1109-L1117.	1.3	57
28	The clinical profile of tuberous sclerosis complex (TSC) in the United Kingdom: A retrospective cohort study in the Clinical Practice Research Datalink (CPRD). European Journal of Paediatric Neurology, 2016, 20, 296-308.	0.7	56
29	Matrix Metalloproteinase-1 Activation Contributes to Airway Smooth Muscle Growth and Asthma Severity. American Journal of Respiratory and Critical Care Medicine, 2017, 195, 1000-1009.	2.5	55
30	Fungal Recognition Enhances Mannose Receptor Shedding through Dectin-1 Engagement. Journal of Biological Chemistry, 2011, 286, 7822-7829.	1.6	53
31	Clinical utility of diagnostic guidelines and putative biomarkers in lymphangioleiomyomatosis. Respiratory Research, 2012, 13, 34.	1.4	50
32	Air travel in women with lymphangioleiomyomatosis. Thorax, 2007, 62, 176-180.	2.7	45
33	Effect of doxycycline on proliferation, MMP production, and adhesion in LAM-related cells. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2010, 299, L393-L400.	1.3	44
34	Tissue and matrix influences on airway smooth muscle function. Pulmonary Pharmacology and Therapeutics, 2009, 22, 379-387.	1.1	40
35	Lymphangioleiomyomatosis. Seminars in Respiratory and Critical Care Medicine, 2002, 23, 085-092.	0.8	36
36	Matrix metalloproteinase-12 (MMP-12) SNP affects MMP activity, lung macrophage infiltration and protects against emphysema in COPD. Thorax, 2011, 66, 970-976.	2.7	36

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37	The Role of Extracellular Matrix Quality in Pulmonary Fibrosis. Respiration, 2014, 88, 487-499.	1.2	36
38	Discoidin domain receptor 1 regulates bronchial epithelial repair and matrix metalloproteinase production. European Respiratory Journal, 2011, 37, 1482-1493.	3.1	35
39	Extra-Cellular Matrix Proteins Induce Matrix Metalloproteinase-1 (MMP-1) Activity and Increase Airway Smooth Muscle Contraction in Asthma. PLoS ONE, 2014, 9, e90565.	1.1	35
40	Use of a three-dimensional cell culture model to study airway smooth muscle-mast cell interactions in airway remodeling. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2009, 296, L1059-L1066.	1.3	34
41	Interstitial lung disease following certolizumab pegol. Rheumatology, 2012, 51, 578-580.	0.9	34
42	MMP-9 protein level does not reflect overall MMP activity in the airways of patients with COPD. Respiratory Medicine, 2008, 102, 845-851.	1.3	33
43	Wild Type Mesenchymal Cells Contribute to the Lung Pathology of Lymphangioleiomyomatosis. PLoS ONE, 2015, 10, e0126025.	1.1	32
44	Natural history of angiomyolipoma in lymphangioleiomyomatosis: implications for screening and surveillance. Orphanet Journal of Rare Diseases, 2014, 9, 151.	1.2	30
45	Phenotypic and functional translation of IL33 genetics in asthma. Journal of Allergy and Clinical Immunology, 2021, 147, 144-157.	1.5	29
46	Levels of circulating MMP-7 degraded elastin are elevated in pulmonary disorders. Clinical Biochemistry, 2015, 48, 1083-1088.	0.8	28
47	Understanding the burden of interstitial lung disease post-COVID-19: the UK Interstitial Lung Disease-Long COVID Study (UKILD-Long COVID). BMJ Open Respiratory Research, 2021, 8, e001049.	1.2	28
48	Phenotypic and functional translation of IL1RL1 locus polymorphisms in lung tissue and asthmatic airway epithelium. JCI Insight, 2020, 5, .	2.3	26
49	Perspectives for improving the evaluation and access of therapies for rare lung diseases in Europe. Respiratory Medicine, 2012, 106, 759-768.	1.3	25
50	The ERS guidelines for LAM: Trying a rationale approach to a rare disease. Respiratory Medicine, 2010, 104, S33-S41.	1.3	23
51	Secretory leukocyte protease inhibitor gene deletion alters bleomycin-induced lung injury, but not development of pulmonary fibrosis. Laboratory Investigation, 2016, 96, 623-631.	1.7	23
52	Cathepsin K in Lymphangioleiomyomatosis. American Journal of Pathology, 2017, 187, 1750-1762.	1.9	23
53	Cardiovascular and inflammatory effects of simvastatin therapy in patients with COPD: a randomized controlled trial. International Journal of COPD, 2015, 10, 211.	0.9	22
54	Analysis of the oestrogen response in an angiomyolipoma derived xenograft model. Endocrine-Related Cancer, 2009, 16, 59-72.	1.6	21

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55	The Role of Inflammation Resolution Speed in Airway Smooth Muscle Mass Accumulation in Asthma: Insight from a Theoretical Model. PLoS ONE, 2014, 9, e90162.	1.1	21
56	The economic burden of tuberous sclerosis complex in the UK: A retrospective cohort study in the Clinical Practice Research Datalink. Journal of Medical Economics, 2016, 19, 1087-1098.	1.0	21
57	Air travel and incidence of pneumothorax in lymphangioleiomyomatosis. Orphanet Journal of Rare Diseases, 2018, 13, 222.	1.2	21
58	TAILS proteomics reveals dynamic changes in airway proteolysis controlling protease activity and innate immunity during COPD exacerbations. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2018, 315, L1003-L1014.	1.3	20
59	A theoretical model of inflammation- and mechanotransduction-driven asthmatic airway remodelling. Biomechanics and Modeling in Mechanobiology, 2018, 17, 1451-1470.	1.4	19
60	Role of the CXCR4/CXCL12 Axis in Lymphangioleiomyomatosis and Angiomyolipoma. Journal of Immunology, 2010, 185, 1812-1821.	0.4	18
61	Matrix metalloproteinases -8 and -9 in the Airways, Blood and Urine During Exacerbations of COPD. COPD: Journal of Chronic Obstructive Pulmonary Disease, 2016, 13, 26-34.	0.7	18
62	Pregnancy in lymphangioleiomyomatosis: clinical and lung function outcomes in two national cohorts. Thorax, 2020, 75, 904-907.	2.7	18
63	The TSC-2 product tuberin is expressed in lymphangioleiomyomatosis and angiomyolipoma. Histopathology, 2002, 40, 458-463.	1.6	16
64	Pulmonary artery aneurysm and tuberous sclerosis. Thorax, 2004, 59, 86-86.	2.7	16
65	In search of the fibrotic epithelial cell: opportunities for a collaborative network. Thorax, 2012, 67, 179-182.	2.7	16
66	The International LAM Registry: A Component of an Innovative Web-Based Clinician, Researcher, and Patient-Driven Rare Disease Research Platform. Lymphatic Research and Biology, 2010, 8, 81-87.	0.5	14
67	Evolution of lung pathology in lymphangioleiomyomatosis: associations with disease course and treatment response. Journal of Pathology: Clinical Research, 2020, 6, 215-226.	1.3	14
68	The vitamin D binding protein axis modifies disease severity in lymphangioleiomyomatosis. European Respiratory Journal, 2018, 52, 1800951.	3.1	13
69	Airway epithelial cell isolation techniques affect DNA methylation profiles with consequences for analysis of asthma related perturbations to DNA methylation. Scientific Reports, 2019, 9, 14409.	1.6	11
70	Cross-sectional study of reversible airway obstruction in LAM: better evidence is needed for bronchodilator and inhaled steroid use. Thorax, 2019, 74, 999-1002.	2.7	11
71	Tuberous Sclerosis Complex (TSC): Expert Recommendations for Provision of Coordinated Care. Frontiers in Neurology, 2019, 10, 1116.	1.1	11
72	Mast-Cell Tryptase Release Contributes to Disease Progression in Lymphangioleiomyomatosis. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 431-444.	2.5	11

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73	Three-Month FVC Change: A Trial Endpoint for Idiopathic Pulmonary Fibrosis Based on Individual Participant Data Meta-analysis. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 936-948.	2.5	11
74	Subcellular distribution of the TSC2 gene product tuberin in human airway smooth muscle cells is driven by multiple localization sequences and is cell-cycle dependent. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2007, 292, L258-L266.	1.3	10
75	Lysyl oxidase like 2 is increased in asthma and contributes to asthmatic airway remodelling. European Respiratory Journal, 2022, 60, 2004361.	3.1	10
76	A 4-year prospective evaluation of protocols to improve clinical outcomes for patients with lymphangioleiomyomatosis in a national clinical centre. Thorax, 2015, 70, 1202-1204.	2.7	9
77	Study of breast cancer incidence in patients of lymphangioleiomyomatosis. Breast Cancer Research and Treatment, 2016, 156, 195-201.	1.1	9
78	Sphingolipid, fatty acid and phospholipid metabolites are associated with disease severity and mTOR inhibition in lymphangioleiomyomatosis. Thorax, 2020, 75, 679-688.	2.7	9
79	<scp>CCR</scp>3 inducedâ€p42/44 <scp>MAPK</scp> activation protects against staurosporine inducedâ€<scp>DNA</scp> fragmentation but not apoptosis in airway smooth muscle cells. Clinical and Experimental Allergy, 2012, 42, 1040-1050.	1.4	8
80	Untangling the protease web in COPD: metalloproteinases in the silent zone. Thorax, 2016, 71, 105-106.	2.7	8
81	Histamine signaling and metabolism identify potential biomarkers and therapies for lymphangioleiomyomatosis. EMBO Molecular Medicine, 2021, 13, e13929.	3.3	6
82	Machine learning can predict disease manifestations and outcomes in lymphangioleiomyomatosis. European Respiratory Journal, 2021, 57, 2003036.	3.1	6
83	Cross talk between LAM cells and fibroblasts may influence alveolar epithelial cell behavior in lymphangioleiomyomatosis. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2022, 322, L283-L293.	1.3	6
84	Living with lymphangioleiomyomatosis. BMJ: British Medical Journal, 2010, 340, c848-c848.	2.4	5
85	Accelerated ¹⁹FÂ·MRI Detection of Matrix Metalloproteinase-2/-9 through Responsive Deactivation of Paramagnetic Relaxation Enhancement. Contrast Media and Molecular Imaging, 2019, 2019, 1-13.	0.4	5
86	COVID-19 in Lymphangioleiomyomatosis. Chest, 2022, 161, 1589-1593.	0.4	5
87	TIMP-1 in asthma: guilty by association. Thorax, 2005, 60, 617-618.	2.7	4
88	An assay to evaluate the long term effects of inflammatory mediators on airway smooth muscle: evidence that TNFÎ± up-regulates 5-HT2A mediated contraction. British Journal of Pharmacology, 2002, 137, 943-944.	2.7	3
89	Doxycycline in lymphangioleiomyomatosis: not all questions are answered. European Respiratory Journal, 2014, 43, 1538-1538.	3.1	3
90	Reply to Yanagisawa: Treatment of Pulmonary Lymphangioleiomyomatosis during Pregnancy. American Journal of Respiratory and Critical Care Medicine, 2018, 197, 1507-1508.	2.5	3

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91	Disease monitoring using lung function trajectory in lymphangioleiomyomatosis: assessment in two national cohorts. Thorax, 2023, 78, 61-68.	2.7	3
92	Lymphangioleiomyomatosis: a case-control study of perinatal and early life events. Thorax, 2003, 58, 979-982.	2.7	2
93	Lymphangioleiomyomatosis. , 2007, , 275-284.		2
94	Pneumothorax and the biology of Birt-Hogg-DubÃ© syndrome. Thorax, 2020, 75, 442-443.	2.7	2
95	Human bronchial epithelial cells from patients with asthma have an altered gene expression profile. ERJ Open Research, 2022, 8, 00625-2021.	1.1	2
96	A survey of use of mTOR inhibitors in patients with lymphangioleiomyomatosis listed for lung transplant. Respiratory Medicine, 2022, 195, 106779.	1.3	1
97	Introduction. Chronic Respiratory Disease, 2005, 2, 73-73.	1.0	0
98	Management of rare diseases in respiratory medicine. Clinical Medicine, 2007, 7, 447-448.	0.8	0
99	Rare Diffuse Interstitial Lung Diseases. , 2012, , 667-675.		0
100	Reply: The ATS/JRS Guidelines on Lymphangioleiomyomatosis: Filling in the Gaps. American Journal of Respiratory and Critical Care Medicine, 2017, 196, 660-661.	2.5	0
101	Mechanisms of Lung Cyst Formation. Respiratory Medicine, 2021, , 21-42.	0.1	0
102	Lymphangioleiomyomatosis. , 2015, , 271-283.		0