Lakshminarayan R Ranganath

List of Publications by Year
in descending order

88 papers	2,254 citations	218677 26 h-index	243625 44 g-index
91 all docs	91 docs citations	91 times ranked	1159 citing authors

#	Article	IF	Citations
1	Metabolomic studies in the inborn error of metabolism alkaptonuria reveal new biotransformations in tyrosine metabolism. Genes and Diseases, 2022, 9, 1129-1142.	3.4	13
2	Identifying joint-specific gait mechanisms causing impaired gait in alkaptonuria patients. Gait and Posture, 2022, 91, 312-317.	1.4	1
3	Comparing nitisinone 2 mg and 10 mg in the treatment of alkaptonuriaâ€"An approach using statistical modelling. JIMD Reports, 2022, 63, 80-92.	1.5	7
4	Effects of a proteinâ€restricted diet on body weight and serum tyrosine concentrations in patients with alkaptonuria. JIMD Reports, 2022, 63, 41-49.	1.5	4
5	Characterization of changes in the tyrosine pathway by 24-h profiling during nitisinone treatment in alkaptonuria. Molecular Genetics and Metabolism Reports, 2022, 30, 100846.	1.1	6
6	Longâ€term low dose nitisinone therapy in adults with alkaptonuria shows no cognitive decline or increased severity of depression. JIMD Reports, 2022, 63, 221-230.	1.5	6
7	Association of alkaptonuria and low dose nitisinone therapy with cataract formation in a large cohort of patients. JIMD Reports, 2022, 63, 351-360.	1.5	6
8	Improving the clinical accuracy and flexibility of the Alkaptonuria severity score index. JIMD Reports, 2022, 63, 361-370.	1.5	3
9	Impact of Nitisinone on the Cerebrospinal Fluid Metabolome of a Murine Model of Alkaptonuria. Metabolites, 2022, 12, 477.	2.9	4
10	Temporal adaptations in the phenylalanine/tyrosine pathway and related factors during nitisinone-induced tyrosinaemia in alkaptonuria. Molecular Genetics and Metabolism, 2022, , .	1.1	6
11	Anatomical Distribution of Ochronotic Pigment in Alkaptonuric Mice is Associated with Calcified Cartilage Chondrocytes at Osteochondral Interfaces. Calcified Tissue International, 2021, 108, 207-218.	3.1	7
12	Expression of tyrosine pathway enzymes in mice demonstrates that homogentisate 1,2â€dioxygenase deficiency in the liver is responsible for homogentisic acidâ€derived ochronotic pigmentation. JIMD Reports, 2021, 58, 52-60.	1.5	3
13	Characterizing the alkaptonuria joint and spine phenotype and assessing the effect of homogentisic acid lowering therapy in a large cohort of 87 patients. Journal of Inherited Metabolic Disease, 2021, 44, 666-676.	3.6	6
14	A case report of pregnancy in untreated alkaptonuria â€" Focus on urinary tissue remodelling markers. Molecular Genetics and Metabolism Reports, 2021, 27, 100766.	1.1	0
15	Development of an effective therapy for alkaptonuria â€" Lessons for osteoarthritis. Rheumatology and Immunology Research, 2021, 2, 79-85.	0.8	3
16	Vitiligo, alkaptonuria, and nitisinoneâ€"A report of three families and review of the literature. JIMD Reports, 2021, 61, 25-33.	1.5	7
17	Evaluating the aortic stenosis phenotype before and after the effect of homogentisic acid lowering therapy: Analysis of a large cohort of eighty-one alkaptonuria patients. Molecular Genetics and Metabolism, 2021, 133, 324-331.	1.1	6
18	â€œLessons from Rare Forms of Osteoarthritisâ€• Calcified Tissue International, 2021, 109, 291-302.	3.1	2

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19	Characterising the arthroplasty in spondyloarthropathy in a large cohort of eightyâ€seven patients with alkaptonuria. Journal of Inherited Metabolic Disease, 2021, 44, 656-665.	3.6	3
20	Treatment of osteoporotic fractures in alkaptonuria by teriparatide stimulates bone formation and decreases fracture rate â€“ A report of two cases. Bone Reports, 2021, 15, 101151.	0.4	3
21	Clinical development innovation in rare diseases: lessons learned and best practices from the DevelopAKUre consortium. Orphanet Journal of Rare Diseases, 2021, 16, 510.	2.7	7
22	Alkaptonuria â€“ Many questions answered, further challenges beckon. Annals of Clinical Biochemistry, 2020, 57, 106-120.	1.6	21
23	Homogentisic acid is not only eliminated by glomerular filtration and tubular secretion but also produced in the kidney in alkaptonuria. Journal of Inherited Metabolic Disease, 2020, 43, 737-747.	3.6	18
24	Dietary restriction of tyrosine and phenylalanine lowers tyrosinemia associated with nitisinone therapy of alkaptonuria. Journal of Inherited Metabolic Disease, 2020, 43, 259-268.	3.6	21
25	The contribution of mouse models in the rare disease alkaptonuria. Drug Discovery Today: Disease Models, 2020, 31, 37-43.	1.2	1
26	Pigmentierungschemie und radikalbasierter Kollagenabbau bei Alkaptonurie und Arthrose. Angewandte Chemie, 2020, 132, 12035-12040.	2.0	0
27	Innentitelbild: Pigmentierungschemie und radikalbasierter Kollagenabbau bei Alkaptonurie und Arthrose (Angew. Chem. 29/2020). Angewandte Chemie, 2020, 132, 11770-11770.	2.0	0
28	Reversal of ochronotic pigmentation in alkaptonuria following nitisinone therapy: Analysis of data from the United Kingdom National Alkaptonuria Centre. JIMD Reports, 2020, 55, 75-87.	1.5	13
29	Fatal acute haemolysis and methaemoglobinaemia in a man with renal failure and Alkaptonuria â€“ Is nitisinone the solution?. Molecular Genetics and Metabolism Reports, 2020, 23, 100588.	1.1	3
30	A patient survey on the impact of alkaptonuria symptoms as perceived by the patients and their experiences of receiving diagnosis and care. JIMD Reports, 2020, 53, 71-79.	1.5	10
31	Pigmentation Chemistry and Radicalâ€Based Collagen Degradation in Alkaptonuria and Osteoarthritic Cartilage. Angewandte Chemie - International Edition, 2020, 59, 11937-11942.	13.8	34
32	Alkaptonuria: Current Perspectives . The Application of Clinical Genetics, 2020, Volume 13, 37-47.	3.0	34
33	Nitisinone causes acquired tyrosinosis in alkaptonuria. Journal of Inherited Metabolic Disease, 2020, 43, 1014-1023.	3.6	20
34	Efficacy and safety of once-daily nitisinone for patients with alkaptonuria (SONIA 2): an international, multicentre, open-label, randomised controlled trial. Lancet Diabetes and Endocrinology,the, 2020, 8, 762-772.	11.4	78
35	Concentric lamellae â€“ novel microanatomical structures in the articular calcified cartilage of mice. Scientific Reports, 2019, 9, 11188.	3.3	5
36	Evaluation of the serum metabolome of patients with alkaptonuria before and after two years of treatment with nitisinone using LCâ€QTOFâ€MS. JIMD Reports, 2019, 48, 67-74.	1.5	11

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37	Ochronotic pigmentation is caused by homogentisic acid and is the key event in alkaptonuria leading to the destructive consequences of the diseaseâ€"A review. Journal of Inherited Metabolic Disease, 2019, 42, 776-792.	3.6	40
38	Conditional targeting in mice reveals that hepatic homogentisate 1,2-dioxygenase activity is essential in reducing circulating homogentisic acid and for effective therapy in the genetic disease alkaptonuria. Human Molecular Genetics, 2019, 28, 3928-3939.	2.9	16
39	Interference of hydroxyphenylpyruvic acid, hydroxyphenyllactic acid and tyrosine on routine serum and urine clinical chemistry assays; implications for biochemical monitoring of patients with alkaptonuria treated with nitisinone. Clinical Biochemistry, 2019, 71, 24-30.	1.9	2
40	Raman Spectroscopy identifies differences in ochronotic and non-ochronotic cartilage; a potential novel technique for monitoring ochronosis. Osteoarthritis and Cartilage, 2019, 27, 1244-1251.	1.3	7
41	Homogentisate 1,2-dioxygenase (HGD) gene variants, their analysis and genotypeâ€"phenotype correlations in the largest cohort of patients with AKU. European Journal of Human Genetics, 2019, 27, 888-902.	2.8	54
42	A Comprehensive LC-QTOF-MS Metabolic Phenotyping Strategy: Application to Alkaptonuria. Clinical Chemistry, 2019, 65, 530-539.	3.2	17
43	Subclinical ochronosis features in alkaptonuria: a cross-sectional study. BMJ Innovations, 2019, 5, 82-91.	1.7	15
44	Alkaptonuria Severity Score Index Revisited: Analysing theÂAKUSSI and Its Subcomponent Features. JIMD Reports, 2018, 41, 53-62.	1.5	9
45	Reduction of frontal plane knee load caused by lateral trunk lean depends on step width. Gait and Posture, 2018, 61, 483-487.	1.4	7
46	Evaluation of the Mitra microsampling device for use with key urinary metabolites in patients with Alkaptonuria. Bioanalysis, 2018, 10, 1919-1932.	1.5	17
47	A new integrated and interactive tool applicable to inborn errors of metabolism: Application to alkaptonuria. Computers in Biology and Medicine, 2018, 103, 1-7.	7.0	17
48	Data on items of AKUSSI in Alkaptonuria collected over three years from the United Kingdom National Alkaptonuria Centre and the impact of nitisinone. Data in Brief, 2018, 20, 1620-1628.	1.0	10
49	Inflammatory and oxidative stress biomarkers in alkaptonuria: data from the DevelopAKUre project. Osteoarthritis and Cartilage, 2018, 26, 1078-1086.	1.3	17
50	Nitisinone arrests ochronosis and decreases rate of progression of Alkaptonuria: Evaluation of the effect of nitisinone in the United Kingdom National Alkaptonuria Centre. Molecular Genetics and Metabolism, 2018, 125, 127-134.	1.1	89
51	Clinical and biochemical assessment of depressive symptoms in patients with Alkaptonuria before and after two years of treatment with nitisinone. Molecular Genetics and Metabolism, 2018, 125, 135-143.	1.1	15
52	Disruption of collagen triple helix hydrogen bonding in ochronotic human cartilage in alkaptonuria observed by dynamic nuclear polarisation-enhanced solid-state nuclear magnetic resonance spectroscopy. Osteoarthritis and Cartilage, 2018, 26, S99.	1.3	1
53	Raman spectroscopy can non-invasively distinguish between ochronotic and non-ochronotic cartilage. Osteoarthritis and Cartilage, 2018, 26, S105.	1.3	2
54	The effect of nitisinone on homogentisic acid and tyrosine: a two-year survey of patients attending the National Alkaptonuria Centre, Liverpool. Annals of Clinical Biochemistry, 2017, 54, 323-330.	1.6	39

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55	Cartilage biomarkers in the osteoarthropathy of alkaptonuria reveal low turnover and accelerated ageing. Rheumatology, 2017, 56, 156-164.	1.9	25
56	Interpreting sources of variation in clinical gait analysis: A case study. Gait and Posture, 2017, 52, 1-4.	1.4	8
57	What does the arthropathy of alkaptonuria teach us about disease mechanisms in osteoarthritis and ageing of joints?. Rheumatology, 2016, 55, 1151-1152.	1.9	6
58	Suitability Of Nitisinone In Alkaptonuria 1 (SONIA 1): an international, multicentre, randomised, open-label, no-treatment controlled, parallel-group, dose-response study to investigate the effect of once daily nitisinone on 24-h urinary homogentisic acid excretion in patients with alkaptonuria after 4â€…weeks of treatment. Annals of the Rheumatic Diseases, 2016, 75, 362-367.	0.9	123
59	Alkaptonuria: An example of a â€œfundamental diseaseâ€â€"A rare disease with important lessons for more common disorders. Seminars in Cell and Developmental Biology, 2016, 52, 53-57.	5.0	36
60	Twelve novel HGD gene variants identified in 99 alkaptonuria patients: focus on â€˜black bone diseaseâ€™ in Italy. European Journal of Human Genetics, 2016, 24, 66-72.	2.8	87
61	Serum markers in alkaptonuria: simultaneous analysis of homogentisic acid, tyrosine and nitisinone by liquid chromatography tandem mass spectrometry. Annals of Clinical Biochemistry, 2015, 52, 597-605.	1.6	46
62	Serum concentrations and urinary excretion of homogentisic acid and tyrosine in normal subjects. Clinical Chemistry and Laboratory Medicine, 2015, 53, e81-3.	2.3	21
63	Lessons from rare diseases of cartilage and bone. Current Opinion in Pharmacology, 2015, 22, 107-114.	3.5	20
64	Relationship Between Serum Concentrations of Nitisinone and Its Effect on Homogentisic Acid and Tyrosine in Patients with Alkaptonuria. JIMD Reports, 2015, 24, 21-27.	1.5	26
65	Fatal oxidative haemolysis and methaemoglobinaemia in a patient with alkaptonuria and acute kidney injury. CKJ: Clinical Kidney Journal, 2015, 8, 109-112.	2.9	10
66	Nitisinone Arrests but Does Not Reverse Ochronosis in Alkaptonuric Mice. JIMD Reports, 2015, 24, 45-50.	1.5	40
67	Diagnostic tools and strategies for assessing disease progression in Alkaptonuria. Expert Opinion on Orphan Drugs, 2015, 3, 705-717.	0.8	1
68	Age-Related Deviation of Gait from Normality in Alkaptonuria. JIMD Reports, 2015, 24, 39-44.	1.5	8
69	192.â€ƒDevelopment of Osteoarthritis in Alkaptonuric Mice. Rheumatology, 2014, 53, i131-i131.	1.9	0
70	Analysis of HGD Gene Mutations in Patients with Alkaptonuria from the United Kingdom: Identification of Novel Mutations. JIMD Reports, 2014, 24, 3-11.	1.5	42
71	The Role of Nitisinone in Tyrosine Pathway Disorders. Current Rheumatology Reports, 2014, 16, 457.	4.7	36
72	Interferences of homogentisic acid (HGA) on routine clinical chemistry assays in serum and urine and the implications for biochemical monitoring of patients with alkaptonuria. Clinical Biochemistry, 2014, 47, 640-647.	1.9	20

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73	Ochronotic osteoarthropathy in a mouse model of alkaptonuria, and its inhibition by nitisinone. Annals of the Rheumatic Diseases, 2014, 73, 284-289.	0.9	88
74	Urine homogentisic acid and tyrosine: Simultaneous analysis by liquid chromatography tandem mass spectrometry. Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences, 2014, 963, 106-112.	2.3	54
75	Acute peripheral administration of synthetic human GLP-1 (7â€“36 amide) decreases circulating IL-6 in obese patients with type 2 diabetes mellitus: A potential role for GLP-1 in modulation of the diabetic pro-inflammatory state?. Regulatory Peptides, 2013, 183, 54-61.	1.9	29
76	Recent advances in management of alkaptonuria (invited review; best practice article). Journal of Clinical Pathology, 2013, 66, 367-373.	2.0	126
77	Efficacy and safety of recombinant human parathyroid hormone (1â€“84) in hypoparathyroidism (REPLACE): a double-blind, placebo-controlled, randomised, phase 3 study. Lancet Diabetes and Endocrinology,the, 2013, 1, 275-283.	11.4	244
78	Ochronosis in a murine model of alkaptonuria is synonymous to that in the human condition. Osteoarthritis and Cartilage, 2012, 20, 880-886.	1.3	45
79	Identification of trabecular excrescences, novel microanatomical structures, present in bone in osteoarthropathies. , 2012, 23, 300-309.		25
80	Living with alkaptonuria. BMJ: British Medical Journal, 2011, 343, d5155-d5155.	2.3	7
81	Cardiovascular manifestations of Alkaptonuria. Journal of Inherited Metabolic Disease, 2011, 34, 1177-1181.	3.6	40
82	A quantitative assessment of alkaptonuria. Journal of Inherited Metabolic Disease, 2011, 34, 1153-1162.	3.6	48
83	Natural history of alkaptonuria revisited: analyses based on scoring systems. Journal of Inherited Metabolic Disease, 2011, 34, 1141-1151.	3.6	69
84	Development of an in vitro model to investigate joint ochronosis in alkaptonuria. Rheumatology, 2011, 50, 271-277.	1.9	62
85	Ultrastructural examination of tissue in a patient with alkaptonuric arthropathy reveals a distinct pattern of binding of ochronotic pigment. Rheumatology, 2010, 49, 1412-1414.	1.9	62
86	Deposition of ochronotic pigment in articular cartilage in alkaptonuria is initiated near the tidemark and progresses to the articular surface. Bone, 2010, 47, S79.	2.9	2
87	The entero-insular axis: implications for human metabolism. Clinical Chemistry and Laboratory Medicine, 2008, 46, 43-56.	2.3	42
88	Method development and validation for analysis of phenylalanine, 4â€hydroxyphenyllactic acid and 4â€hydroxyphenylpyruvic acid in serum and urine. JIMD Reports, 0, , .	1.5	6