Terry G J Derks

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69 968 19 27 g-index

78 1,237 5.4 4.2 ext. papers ext. citations avg, IF L-index

#	Paper	IF	Citations
69	Glycogen storage disease type III: diagnosis, genotype, management, clinical course and outcome. <i>Journal of Inherited Metabolic Disease</i> , 2016 , 39, 697-704	5.4	83
68	The natural history of medium-chain acyl CoA dehydrogenase deficiency in the Netherlands: clinical presentation and outcome. <i>Journal of Pediatrics</i> , 2006 , 148, 665-670	3.6	77
67	Treating neutropenia and neutrophil dysfunction in glycogen storage disease type Ib with an SGLT2 inhibitor. <i>Blood</i> , 2020 , 136, 1033-1043	2.2	39
66	Inborn Errors of Metabolism with Hypoglycemia: Glycogen Storage Diseases and Inherited Disorders of Gluconeogenesis. <i>Pediatric Clinics of North America</i> , 2018 , 65, 247-265	3.6	36
65	Evaluation of glycogen storage disease as a cause of ketotic hypoglycemia in children. <i>Journal of Inherited Metabolic Disease</i> , 2015 , 38, 489-93	5.4	34
64	Lipids in hepatic glycogen storage diseases: pathophysiology, monitoring of dietary management and future directions. <i>Journal of Inherited Metabolic Disease</i> , 2015 , 38, 537-43	5.4	34
63	Disturbed hepatic carbohydrate management during high metabolic demand in medium-chain acyl-CoA dehydrogenase (MCAD)-deficient mice. <i>Hepatology</i> , 2008 , 47, 1894-904	11.2	34
62	Dietary management in glycogen storage disease type III: what is the evidence?. <i>Journal of Inherited Metabolic Disease</i> , 2015 , 38, 545-50	5.4	30
61	Safe and unsafe duration of fasting for children with MCAD deficiency. <i>European Journal of Pediatrics</i> , 2007 , 166, 5-11	4.1	29
60	Inhibition of mitochondrial fatty acid oxidation in vivo only slightly suppresses gluconeogenesis but enhances clearance of glucose in mice. <i>Hepatology</i> , 2008 , 47, 1032-42	11.2	26
59	The difference between observed and expected prevalence of MCAD deficiency in The Netherlands: a genetic epidemiological study. <i>European Journal of Human Genetics</i> , 2005 , 13, 947-52	5.3	26
58	Inborn Errors of Metabolism That Cause Sudden Infant Death: A Systematic Review with Implications for Population Neonatal Screening Programmes. <i>Neonatology</i> , 2016 , 109, 297-302	4	25
57	Pathogenic variants in glutamyl-tRNA amidotransferase subunits cause a lethal mitochondrial cardiomyopathy disorder. <i>Nature Communications</i> , 2018 , 9, 4065	17.4	24
56	Dietary treatment of glycogen storage disease type Ia: uncooked cornstarch and/or continuous nocturnal gastric drip-feeding?. <i>Molecular Genetics and Metabolism</i> , 2013 , 109, 1-2	3.7	23
55	Nine years of newborn screening for classical galactosemia in the Netherlands: Effectiveness of screening methods, and identification of patients with previously unreported phenotypes. <i>Molecular Genetics and Metabolism</i> , 2017 , 120, 223-228	3.7	21
54	Risk stratification by residual enzyme activity after newborn screening for medium-chain acyl-CoA dehyrogenase deficiency: data from a cohort study. <i>Orphanet Journal of Rare Diseases</i> , 2012 , 7, 30	4.2	20
53	Cost-effectiveness of neonatal screening for medium chain acyl-CoA dehydrogenase deficiency: the homogeneous population of The Netherlands. <i>Journal of Pediatrics</i> , 2007 , 151, 115-20, 120.e1-3	3.6	19

(2017-2019)

52	Movement disorders and nonmotor neuropsychological symptoms in children and adults with classical galactosemia. <i>Journal of Inherited Metabolic Disease</i> , 2019 , 42, 451-458	5.4	19	
51	Impact of newborn screening for very-long-chain acyl-CoA dehydrogenase deficiency on genetic, enzymatic, and clinical outcomes. <i>Journal of Inherited Metabolic Disease</i> , 2019 , 42, 414-423	5.4	19	
50	Research priorities for liver glycogen storage disease: An international priority setting partnership with the James Lind Alliance. <i>Journal of Inherited Metabolic Disease</i> , 2020 , 43, 279-289	5.4	19	
49	Proposal for an individualized dietary strategy in patients with very long-chain acyl-CoA dehydrogenase deficiency. <i>Journal of Inherited Metabolic Disease</i> , 2019 , 42, 159-168	5.4	18	
48	Favorable outcome after physiologic dose of sodium-D,L-3-hydroxybutyrate in severe MADD. <i>Pediatrics</i> , 2014 , 134, e1224-8	7.4	17	
47	Experimental evidence for protein oxidative damage and altered antioxidant defense in patients with medium-chain acyl-CoA dehydrogenase deficiency. <i>Journal of Inherited Metabolic Disease</i> , 2014 , 37, 783-9	5.4	16	
46	Clinical and biochemical heterogeneity between patients with glycogen storage disease type IA: the added value of CUSUM for metabolic control. <i>Journal of Inherited Metabolic Disease</i> , 2017 , 40, 695-	76̄2 ⁴	15	
45	Living on the edge: substrate competition explains loss of robustness in mitochondrial fatty-acid oxidation disorders. <i>BMC Biology</i> , 2016 , 14, 107	7-3	15	
44	Biallelic variants in WARS2 encoding mitochondrial tryptophanyl-tRNA synthase in six individuals with mitochondrial encephalopathy. <i>Human Mutation</i> , 2017 , 38, 1786-1795	4.7	15	
43	In vitro digestion of starches in a dynamic gastrointestinal model: an innovative study to optimize dietary management of patients with hepatic glycogen storage diseases. <i>Journal of Inherited Metabolic Disease</i> , 2015 , 38, 529-36	5.4	14	
42	Efficacy and safety of D,L-3-hydroxybutyrate (D,L-3-HB) treatment in multiple acyl-CoA dehydrogenase deficiency. <i>Genetics in Medicine</i> , 2020 , 22, 908-916	8.1	13	
41	A nationwide retrospective observational study of population newborn screening for medium-chain acyl-CoA dehydrogenase (MCAD) deficiency in the Netherlands. <i>Journal of Inherited Metabolic Disease</i> , 2019 , 42, 890-897	5.4	12	
40	Normoglycemic Ketonemia as Biochemical Presentation in Ketotic Glycogen Storage Disease. <i>JIMD Reports</i> , 2016 , 28, 41-47	1.9	12	
39	Dietary lipids in glycogen storage disease type III: A systematic literature study, case studies, and future recommendations. <i>Journal of Inherited Metabolic Disease</i> , 2020 , 43, 770-777	5.4	12	
38	Hepatocytes contribute to residual glucose production in a mouse model for glycogen storage disease type Ia. <i>Hepatology</i> , 2017 , 66, 2042-2054	11.2	12	
37	Determination of amylose/amylopectin ratio of starches. <i>Journal of Inherited Metabolic Disease</i> , 2015 , 38, 985-6	5.4	11	
36	Muscle Ultrasound in Patients with Glycogen Storage Disease Types I and III. <i>Ultrasound in Medicine and Biology</i> , 2016 , 42, 133-42	3.5	11	
35	Inflammatory Bowel Disease in Glycogen Storage Disease Type Ia. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2017 , 64, e52-e54	2.8	10	

34	Safety issues associated with dietary management in patients with hepatic glycogen storage disease. <i>Molecular Genetics and Metabolism</i> , 2018 , 125, 79-85	3.7	10
33	Prediction of disease severity in multiple acyl-CoA dehydrogenase deficiency: A retrospective and laboratory cohort study. <i>Journal of Inherited Metabolic Disease</i> , 2019 , 42, 878-889	5.4	10
32	Recombinant phenylalanine ammonia lyase in phenylketonuria. <i>Lancet, The</i> , 2014 , 384, 6-8	40	10
31	Improved inflammatory bowel disease, wound healing and normal oxidative burst under treatment with empagliflozin in glycogen storage disease type lb. <i>Orphanet Journal of Rare Diseases</i> , 2020 , 15, 218	4.2	10
30	Impaired Very-Low-Density Lipoprotein catabolism links hypoglycemia to hypertriglyceridemia in Glycogen Storage Disease type[la. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 879-892	5.4	7
29	What Is the Best Blood Sampling Time for Metabolic Control of Phenylalanine and Tyrosine Concentrations in Tyrosinemia Type 1 Patients?. <i>JIMD Reports</i> , 2017 , 36, 49-57	1.9	6
28	A preliminary study of telemedicine for patients with hepatic glycogen storage disease and their healthcare providers: from bedside to home site monitoring. <i>Journal of Inherited Metabolic Disease</i> , 2018 , 41, 929-936	5.4	6
27	Complex patterns of inheritance, including synergistic heterozygosity, in inborn errors of metabolism: Implications for precision medicine driven diagnosis and treatment. <i>Molecular Genetics and Metabolism</i> , 2019 , 128, 1-9	3.7	6
26	Crohn disease-like enterocolitis remission after empagliflozin treatment in a child with glycogen storage disease type Ib: a case report. <i>Italian Journal of Pediatrics</i> , 2021 , 47, 149	3.2	6
25	Glycogen storage disease type Ia: Adult presentation with microcytic anemia and liver adenomas. <i>Hepatology</i> , 2018 , 68, 780-782	11.2	5
24	Acute exercise in treated phenylketonuria patients: Physical activity and biochemical response. <i>Molecular Genetics and Metabolism Reports</i> , 2015 , 5, 55-59	1.8	5
23	The multiple faces of urinary glucose tetrasaccharide as biomarker for patients with hepatic glycogen storage diseases. <i>Genetics in Medicine</i> , 2020 , 22, 1915-1916	8.1	5
22	The potential of dietary treatment in patients with glycogen storage disease type IV. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 693-704	5.4	4
21	Glycogen storage disease type 1a is associated with disturbed vitamin A metabolism and elevated serum retinol levels. <i>Human Molecular Genetics</i> , 2020 , 29, 264-273	5.6	4
20	A generic emergency protocol for patients with inborn errors of metabolism causing fasting intolerance: A retrospective, single-center study and the generation of www.emergencyprotocol.net. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 1124-1135	5.4	4
19	Changes in pediatric plasma acylcarnitines upon fasting for refined interpretation of metabolic stress. <i>Molecular Genetics and Metabolism</i> , 2019 , 127, 327-335	3.7	3
18	Hepatic Glycogen Storage Diseases: Toward One Global Collaborative Network. <i>FIRE Forum for International Research in Education</i> , 2017 , 5, 232640981773300	1.4	3
17	A retrospective in-depth analysis of continuous glucose monitoring datasets for patients with hepatic glycogen storage disease: Recommended outcome parameters for glucose management. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 1136-1150	5.4	3

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16	Effects of acute nutritional ketosis during exercise in adults with glycogen storage disease type IIIa are phenotype-specific: An investigator-initiated, randomized, crossover study. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 226-239	5.4	3
15	Enantiomer-specific pharmacokinetics of D,L-3-hydroxybutyrate: Implications for the treatment of multiple acyl-CoA dehydrogenase deficiency. <i>Journal of Inherited Metabolic Disease</i> , 2021 , 44, 926-938	5.4	3
14	Neonates at risk of medium-chain acyl-CoA dehydrogenase deficiency: a perinatal protocol for use before population neonatal screening test results become available. <i>Genetics in Medicine</i> , 2016 , 18, 132	.2 ⁸ 1 ¹ 32	3 ²
13	Transcriptome analysis suggests a compensatory role of the cofactors coenzyme A and NAD in medium-chain acyl-CoA dehydrogenase knockout mice. <i>Scientific Reports</i> , 2019 , 9, 14539	4.9	2
12	Glycogen Storage Disease Type Ia: Current Management Options, Burden and Unmet Needs. <i>Nutrients</i> , 2021 , 13,	6.7	2
11	Proposal for an individualized dietary strategy in patients with very long-chain acyl-CoA dehydrogenase deficiency 2019 , 42, 159		2
10	A triple-blinded crossover study to evaluate the short-term safety of sweet manioc starch for the treatment of glycogen storage disease type Ia. <i>Orphanet Journal of Rare Diseases</i> , 2021 , 16, 254	4.2	2
9	Efficacy and safety of empagliflozin in glycogen storage disease type Ib: Data from an international questionnaire <i>Genetics in Medicine</i> , 2022 ,	8.1	2
8	Instability of Acylcarnitines in Stored Dried Blood Spots: The Impact on Retrospective Analysis of Biomarkers for Inborn Errors of Metabolism. <i>International Journal of Neonatal Screening</i> , 2020 , 6,	2.6	1
7	A retrospective study of eating and psychosocial problems in patients with hepatic glycogen storage diseases and idiopathic ketotic hypoglycemia: Towards a standard set of patient-reported outcome measures <i>JIMD Reports</i> , 2022 , 63, 29-40	1.9	1
6	Orthotopic Liver Transplantation in Glycogen Storage Disease Type 1a: Perioperative Glucose and Lactate Homeostasis. <i>FIRE Forum for International Research in Education</i> , 2016 , 4, 232640981664959	1.4	1
5	Modeling Phenotypic Heterogeneity of Glycogen Storage Disease Type 1a Liver Disease in Mice by Somatic CRISPR/CRISPR-associated protein 9-Mediated Gene Editing. <i>Hepatology</i> , 2021 , 74, 2491-2507	11.2	O
4	Neonatal Long-Chain 3-Ketoacyl-CoA Thiolase deficiency: Clinical-biochemical phenotype, sodium-D,L-3-hydroxybutyrate treatment experience and cardiac evaluation using speckle echocardiography. <i>Molecular Genetics and Metabolism Reports</i> , 2022 , 31, 100873	1.8	O
3	High childhood serum triglyceride concentrations associate with hepatocellular adenoma development in patients with glycogen storage disease type Ia. <i>JHEP Reports</i> , 2022 , 100512	10.3	0
2	Glycogen Storage Liver Diseases 2020 , 749-754		
1	Disorders of Carbohydrate Absorption, Transmembrane Transport and Metabolism 2022 , 649-700		