Terry G J Derks

List of Publications by Year in Descending Order

69 papers	968 citations	19 h-index	27 g-index
78 ext. papers	1,237 ext. citations	5.4 avg, IF	4.2 L-index

#	Paper	IF	Citations
69	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.. *JIMD Reports*, **2022**, 63, 29-40	1.9	1
68	Disorders of Carbohydrate Absorption, Transmembrane Transport and Metabolism **2022**, 649-700		
67	Efficacy and safety of empagliflozin in glycogen storage disease type Ib: Data from an international questionnaire.. *Genetics in Medicine*, **2022**,	8.1	2
66	Neonatal Long-Chain 3-Ketoacyl-CoA Thiolase deficiency: Clinical-biochemical phenotype, sodium-D,L-3-hydroxybutyrate treatment experience and cardiac evaluation using speckle echocardiography. *Molecular Genetics and Metabolism Reports*, **2022**, 31, 100873	1.8	0
65	High childhood serum triglyceride concentrations associate with hepatocellular adenoma development in patients with glycogen storage disease type Ia. *JHEP Reports*, **2022**, 100512	10.3	0
64	Glycogen Storage Disease Type Ia: Current Management Options, Burden and Unmet Needs. *Nutrients*, **2021**, 13,	6.7	2
63	The potential of dietary treatment in patients with glycogen storage disease type IV. *Journal of Inherited Metabolic Disease*, **2021**, 44, 693-704	5.4	4
62	Impaired Very-Low-Density Lipoprotein catabolism links hypoglycemia to hypertriglyceridemia in Glycogen Storage Disease type Ia. *Journal of Inherited Metabolic Disease*, **2021**, 44, 879-892	5.4	7
61	A retrospective in-depth analysis of continuous glucose monitoring datasets for patients with hepatic glycogen storage disease: Recommended outcome parameters for glucose management. *Journal of Inherited Metabolic Disease*, **2021**, 44, 1136-1150	5.4	3
60	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. *Journal of Inherited Metabolic Disease*, **2021**, 44, 1124-1135	5.4	4
59	A triple-blinded crossover study to evaluate the short-term safety of sweet manioc starch for the treatment of glycogen storage disease type Ia. *Orphanet Journal of Rare Diseases*, **2021**, 16, 254	4.2	2
58	Effects of acute nutritional ketosis during exercise in adults with glycogen storage disease type IIIa are phenotype-specific: An investigator-initiated, randomized, crossover study. *Journal of Inherited Metabolic Disease*, **2021**, 44, 226-239	5.4	3
57	Enantiomer-specific pharmacokinetics of D,L-3-hydroxybutyrate: Implications for the treatment of multiple acyl-CoA dehydrogenase deficiency. *Journal of Inherited Metabolic Disease*, **2021**, 44, 926-938	5.4	3
56	Crohn disease-like enterocolitis remission after empagliflozin treatment in a child with glycogen storage disease type Ib: a case report. *Italian Journal of Pediatrics*, **2021**, 47, 149	3.2	6
55	Modeling Phenotypic Heterogeneity of Glycogen Storage Disease Type 1a Liver Disease in Mice by Somatic CRISPR/CRISPR-associated protein 9-Mediated Gene Editing. *Hepatology*, **2021**, 74, 2491-2507	11.2	0
54	Instability of Acylcarnitines in Stored Dried Blood Spots: The Impact on Retrospective Analysis of Biomarkers for Inborn Errors of Metabolism. *International Journal of Neonatal Screening*, **2020**, 6,	2.6	1
53	Treating neutropenia and neutrophil dysfunction in glycogen storage disease type Ib with an SGLT2 inhibitor. *Blood*, **2020**, 136, 1033-1043	2.2	39

52	Dietary lipids in glycogen storage disease type III: A systematic literature study, case studies, and future recommendations. *Journal of Inherited Metabolic Disease*, **2020**, 43, 770-777	5.4	12
51	Glycogen Storage Liver Diseases **2020**, 749-754		
50	Efficacy and safety of D,L-3-hydroxybutyrate (D,L-3-HB) treatment in multiple acyl-CoA dehydrogenase deficiency. *Genetics in Medicine*, **2020**, 22, 908-916	8.1	13
49	Glycogen storage disease type 1a is associated with disturbed vitamin A metabolism and elevated serum retinol levels. *Human Molecular Genetics*, **2020**, 29, 264-273	5.6	4
48	The multiple faces of urinary glucose tetrasaccharide as biomarker for patients with hepatic glycogen storage diseases. *Genetics in Medicine*, **2020**, 22, 1915-1916	8.1	5
47	Improved inflammatory bowel disease, wound healing and normal oxidative burst under treatment with empagliflozin in glycogen storage disease type Ib. *Orphanet Journal of Rare Diseases*, **2020**, 15, 218	4.2	10
46	Research priorities for liver glycogen storage disease: An international priority setting partnership with the James Lind Alliance. *Journal of Inherited Metabolic Disease*, **2020**, 43, 279-289	5.4	19
45	A nationwide retrospective observational study of population newborn screening for medium-chain acyl-CoA dehydrogenase (MCAD) deficiency in the Netherlands. *Journal of Inherited Metabolic Disease*, **2019**, 42, 890-897	5.4	12
44	Proposal for an individualized dietary strategy in patients with very long-chain acyl-CoA dehydrogenase deficiency. *Journal of Inherited Metabolic Disease*, **2019**, 42, 159-168	5.4	18
43	Complex patterns of inheritance, including synergistic heterozygosity, in inborn errors of metabolism: Implications for precision medicine driven diagnosis and treatment. *Molecular Genetics and Metabolism*, **2019**, 128, 1-9	3.7	6
42	Changes in pediatric plasma acylcarnitines upon fasting for refined interpretation of metabolic stress. *Molecular Genetics and Metabolism*, **2019**, 127, 327-335	3.7	3
41	Prediction of disease severity in multiple acyl-CoA dehydrogenase deficiency: A retrospective and laboratory cohort study. *Journal of Inherited Metabolic Disease*, **2019**, 42, 878-889	5.4	10
40	Transcriptome analysis suggests a compensatory role of the cofactors coenzyme A and NAD in medium-chain acyl-CoA dehydrogenase knockout mice. *Scientific Reports*, **2019**, 9, 14539	4.9	2
39	Proposal for an individualized dietary strategy in patients with very long-chain acyl-CoA dehydrogenase deficiency **2019**, 42, 159		2
38	Movement disorders and nonmotor neuropsychological symptoms in children and adults with classical galactosemia. *Journal of Inherited Metabolic Disease*, **2019**, 42, 451-458	5.4	19
37	Impact of newborn screening for very-long-chain acyl-CoA dehydrogenase deficiency on genetic, enzymatic, and clinical outcomes. *Journal of Inherited Metabolic Disease*, **2019**, 42, 414-423	5.4	19
36	Glycogen storage disease type Ia: Adult presentation with microcytic anemia and liver adenomas. *Hepatology*, **2018**, 68, 780-782	11.2	5
35	A preliminary study of telemedicine for patients with hepatic glycogen storage disease and their healthcare providers: from bedside to home site monitoring. *Journal of Inherited Metabolic Disease*, **2018**, 41, 929-936	5.4	6

34	Inborn Errors of Metabolism with Hypoglycemia: Glycogen Storage Diseases and Inherited Disorders of Gluconeogenesis. *Pediatric Clinics of North America*, **2018**, 65, 247-265	3.6	36
33	Safety issues associated with dietary management in patients with hepatic glycogen storage disease. *Molecular Genetics and Metabolism*, **2018**, 125, 79-85	3.7	10
32	Pathogenic variants in glutamyl-tRNA amidotransferase subunits cause a lethal mitochondrial cardiomyopathy disorder. *Nature Communications*, **2018**, 9, 4065	17.4	24
31	Inflammatory Bowel Disease in Glycogen Storage Disease Type Ia. *Journal of Pediatric Gastroenterology and Nutrition*, **2017**, 64, e52-e54	2.8	10
30	Nine years of newborn screening for classical galactosemia in the Netherlands: Effectiveness of screening methods, and identification of patients with previously unreported phenotypes. *Molecular Genetics and Metabolism*, **2017**, 120, 223-228	3.7	21
29	What Is the Best Blood Sampling Time for Metabolic Control of Phenylalanine and Tyrosine Concentrations in Tyrosinemia Type 1 Patients?. *JIMD Reports*, **2017**, 36, 49-57	1.9	6
28	Clinical and biochemical heterogeneity between patients with glycogen storage disease type IA: the added value of CUSUM for metabolic control. *Journal of Inherited Metabolic Disease*, **2017**, 40, 695-702	5.4	15
27	Biallelic variants in WARS2 encoding mitochondrial tryptophanyl-tRNA synthase in six individuals with mitochondrial encephalopathy. *Human Mutation*, **2017**, 38, 1786-1795	4.7	15
26	Hepatocytes contribute to residual glucose production in a mouse model for glycogen storage disease type Ia. *Hepatology*, **2017**, 66, 2042-2054	11.2	12
25	Hepatic Glycogen Storage Diseases: Toward One Global Collaborative Network. *FIRE Forum for International Research in Education*, **2017**, 5, 232640981773300	1.4	3
24	Normoglycemic Ketonemia as Biochemical Presentation in Ketotic Glycogen Storage Disease. *JIMD Reports*, **2016**, 28, 41-47	1.9	12
23	Neonates at risk of medium-chain acyl-CoA dehydrogenase deficiency: a perinatal protocol for use before population neonatal screening test results become available. *Genetics in Medicine*, **2016**, 18, 1322-1323	8.1	2
22	Living on the edge: substrate competition explains loss of robustness in mitochondrial fatty-acid oxidation disorders. *BMC Biology*, **2016**, 14, 107	7.3	15
21	Inborn Errors of Metabolism That Cause Sudden Infant Death: A Systematic Review with Implications for Population Neonatal Screening Programmes. *Neonatology*, **2016**, 109, 297-302	4	25
20	Muscle Ultrasound in Patients with Glycogen Storage Disease Types I and III. *Ultrasound in Medicine and Biology*, **2016**, 42, 133-42	3.5	11
19	Orthotopic Liver Transplantation in Glycogen Storage Disease Type 1a: Perioperative Glucose and Lactate Homeostasis. *FIRE Forum for International Research in Education*, **2016**, 4, 232640981664959	1.4	1
18	Glycogen storage disease type III: diagnosis, genotype, management, clinical course and outcome. *Journal of Inherited Metabolic Disease*, **2016**, 39, 697-704	5.4	83
17	Evaluation of glycogen storage disease as a cause of ketotic hypoglycemia in children. *Journal of Inherited Metabolic Disease*, **2015**, 38, 489-93	5.4	34

16	Determination of amylose/amylopectin ratio of starches. *Journal of Inherited Metabolic Disease*, **2015**, 38, 985-6	5.4	11
15	Lipids in hepatic glycogen storage diseases: pathophysiology, monitoring of dietary management and future directions. *Journal of Inherited Metabolic Disease*, **2015**, 38, 537-43	5.4	34
14	Dietary management in glycogen storage disease type III: what is the evidence?. *Journal of Inherited Metabolic Disease*, **2015**, 38, 545-50	5.4	30
13	Acute exercise in treated phenylketonuria patients: Physical activity and biochemical response. *Molecular Genetics and Metabolism Reports*, **2015**, 5, 55-59	1.8	5
12	In vitro digestion of starches in a dynamic gastrointestinal model: an innovative study to optimize dietary management of patients with hepatic glycogen storage diseases. *Journal of Inherited Metabolic Disease*, **2015**, 38, 529-36	5.4	14
11	Recombinant phenylalanine ammonia lyase in phenylketonuria. *Lancet, The*, **2014**, 384, 6-8	40	10
10	Experimental evidence for protein oxidative damage and altered antioxidant defense in patients with medium-chain acyl-CoA dehydrogenase deficiency. *Journal of Inherited Metabolic Disease*, **2014** , 37, 783-9	5.4	16
9	Favorable outcome after physiologic dose of sodium-D,L-3-hydroxybutyrate in severe MADD. *Pediatrics*, **2014**, 134, e1224-8	7.4	17
8	Dietary treatment of glycogen storage disease type Ia: uncooked cornstarch and/or continuous nocturnal gastric drip-feeding?. *Molecular Genetics and Metabolism*, **2013**, 109, 1-2	3.7	23
7	Risk stratification by residual enzyme activity after newborn screening for medium-chain acyl-CoA dehyrogenase deficiency: data from a cohort study. *Orphanet Journal of Rare Diseases*, **2012**, 7, 30	4.2	20
6	Inhibition of mitochondrial fatty acid oxidation in vivo only slightly suppresses gluconeogenesis but enhances clearance of glucose in mice. *Hepatology*, **2008**, 47, 1032-42	11.2	26
5	Disturbed hepatic carbohydrate management during high metabolic demand in medium-chain acyl-CoA dehydrogenase (MCAD)-deficient mice. *Hepatology*, **2008**, 47, 1894-904	11.2	34
4	Cost-effectiveness of neonatal screening for medium chain acyl-CoA dehydrogenase deficiency: the homogeneous population of The Netherlands. *Journal of Pediatrics*, **2007**, 151, 115-20, 120.e1-3	3.6	19
3	Safe and unsafe duration of fasting for children with MCAD deficiency. *European Journal of Pediatrics*, **2007**, 166, 5-11	4.1	29
2	The natural history of medium-chain acyl CoA dehydrogenase deficiency in the Netherlands: clinical presentation and outcome. *Journal of Pediatrics*, **2006**, 148, 665-670	3.6	77
1	The difference between observed and expected prevalence of MCAD deficiency in The Netherlands: a genetic epidemiological study. *European Journal of Human Genetics*, **2005**, 13, 947-52	5.3	26