

Terry G. J. Derks, MD, PhD
consultant pediatric metabolic diseases

University of Groningen, UMC Groningen
Beatrix Children's Hospital, section of metabolic diseases
Hanzeplein 1
9713 GZ Groningen, the Netherlands



Email: t.g.j.derks@umcg.nl
Phone: +31 50 361 4147

Research output

Modeling phenotypic heterogeneity of Glycogen Storage Disease type Ia liver disease in mice by somatic CRISPR/Cas9-mediated gene editing

Rutten, M. G. S., Derks, T. G. J., Huijkman, N. C. A., Bos, T., Kloosterhuis, N. J., van de Kolk, K. C. W. A., Wolters, J. C., Koster, M. H., Bongiovanni, L., Thomas, R. E., de Bruin, A., van de Sluis, B. & Oosterveer, M. H., 22-Jun-2021, (E-pub ahead of print) In: *Hepatology*. p. 1-37 37 p.

A triple-blinded crossover study to evaluate the short-term safety of sweet manioc starch for the treatment of glycogen storage disease type Ia

Monteiro, V. C. L., de Oliveira, B. M., Dos Santos, B. B., Sperb-Ludwig, F., Refosco, L. F., Nalin, T., Derks, T. G. J., Moura de Souza, C. F. & Schwartz, I. V. D., 3-Jun-2021, In: *Orphanet journal of rare diseases*. 16, 1, 10 p., 254.

A retrospective in-depth analysis of continuous glucose monitoring datasets for patients with hepatic glycogen storage disease: Recommended outcome parameters for glucose management

Peeks, F., Hoogeveen, I. J., Feldbrugge, R. L., Burghard, R., de Boer, F., Fokkert-Wilts, M. J., van der Klauw, M. M., Oosterveer, M. H. & Derks, T. G. J., 5-May-2021, (E-pub ahead of print) In: *Journal of Inherited Metabolic Disease*. 15 p.

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

CONNECT MetabERN collaboration group, Rossi, A., Hoogeveen, I. J., Lubout, C. M. A., de Boer, F., Fokkert-Wilts, M. J., Rodenburg, I. L., van Dam, E., Grünert, S. C., Martinelli, D., Scarpa, M., Dekker, H., Te Boekhorst, S. T., van Spronsen, F. J. & Derks, T. G. J., 4-May-2021, (E-pub ahead of print) In: *Journal of Inherited Metabolic Disease*. 12 p.

Impaired Very-Low-Density Lipoprotein catabolism links hypoglycemia to hypertriglyceridemia in Glycogen Storage Disease type Ia

Hoogerland, J. A., Peeks, F., Hijmans, B. S., Wolters, J. C., Kooijman, S., Bos, T., Bleeker, A., van Dijk, T. H., Wolters, H., Gerding, A., van Eunen, K., Havinga, R., Pronk, A. C. M., Rensen, P. C. N., Mithieux, G., Rajas, F., Kuipers, F., Reijngoud, D-J., Derks, T. G. J. & Oosterveer, M. H., 7-Apr-2021, (E-pub ahead of print) In: *Journal of Inherited Metabolic Disease*. 14 p.

Experimentele gentherapie voor patiënten met zeer zeldzame erfelijke ziekte

Derks, TG., 8-Feb-2021

Enantiomer-specific pharmacokinetics of D,L-3-hydroxybutyrate: Implications for the treatment of multiple acyl-CoA dehydrogenase deficiency

van Rijt, W. J., Van Hove, J. L. K., Vaz, F. M., Havinga, R., Allersma, D. P., Zijp, T. R., Bedoyan, J. K., Heiner-Fokkema, M. R., Reijngoud, D-J., Geraghty, M. T., Wanders, R. J. A., Oosterveer, M. H. & Derks, T. G. J., 5-Feb-2021, (E-pub ahead of print) In: *Journal of Inherited Metabolic Disease*. 13 p.

Effects of acute nutritional ketosis during exercise in adults with glycogen storage disease type IIIa: An investigator-initiated, randomized, crossover study

Hoogeveen, I. J., de Boer, F., Boonstra, W. F., van Der Schaaf, C. J., Steuerwald, U., Sibeijn-Kuiper, A. J., Vegter, R. J. K., van Der Hoeven, J. H., Heiner-Fokkema, M. R., Clarke, K. C., Cox, P. J., Derks, T. G. J. & Jeneson, J. A. L., Jan-2021, In: *Journal of Inherited Metabolic Disease*. 44, 1, p. 226-239 14 p.

The potential of dietary treatment in patients with glycogen storage disease type IV

Derks, T. G. J., Peeks, F., de Boer, F., Fokkert-Wilts, M., van der Doef, H. P. J., van den Heuvel, M. C., Szymańska, E., Rokicki, D., Ryan, P. T. & Weinstein, D. A., 21-Dec-2020, In: *Journal of Inherited Metabolic Disease*. 12 p.

Instability of Acylcarnitines in Stored Dried Blood Spots: The Impact on Retrospective Analysis of Biomarkers for Inborn Errors of Metabolism

van Rijt, W. J., Schielen, P. C. J. I., Özer, Y., Bijsterveld, K., van der Sluijs, F. H., Derks, T. G. J. & Heiner-Fokkema, M. R., Dec-2020, In: International journal of neonatal screening. 6, 4, 13 p., 83.

Complex patterns of inheritance, including synergistic heterozygosity, in inborn errors of metabolism: Implications for precision medicine driven diagnosis and treatment

Vockley, J., Dobrowolski, S. F., Arnold, G. L., Guerrero, R. B., Derks, T. G. J. & Weinstein, D. A., Sep-2020, In: Molecular Genetics and Metabolism. 128, 1-2, p. 1-9 9 p.

Treating neutropenia and neutrophil dysfunction in glycogen storage disease IB with an SGLT2-inhibitor

Wortmann, S. B., Van Hove, J. L. K., Derks, T. G. J., Chevalier, N., Knight, V., Koller, A., Oussuren, E., Mayr, J. A., van Spronsen, F. J., Lagler, F. B., Gaughan, S., Van Schaftingen, E. & Veiga-da-Cunha, M., 27-Aug-2020, In: Blood. 136, 9, p. 1033-1043 11 p.

Improved inflammatory bowel disease, wound healing and normal oxidative burst under treatment with empagliflozin in glycogen storage disease type Ib

Grünert, S. C., Elling, R., Maag, B., Wortmann, S. B., Derks, T. G. J., Hannibal, L., Schumann, A., Rosenbaum-Fabian, S. & Spiekerkoetter, U., 24-Aug-2020, In: Orphanet journal of rare diseases. 15, 1, 8 p., 218.

The multiple faces of urinary glucose tetrasaccharide as biomarker for patients with hepatic glycogen storage diseases

Heiner-Fokkema, M. R., van der Krogt, J., de Boer, F., Fokkert-Wilts, M. J., Maatman, R. G. H. J., Hoogeveen, I. J. & Derks, T. G. J., 13-Jul-2020, (E-pub ahead of print) In: Genetics in Medicine. 11, p. 1915-1916 2 p.

Dietary lipids in glycogen storage disease type III: a systematic literature study, case studies and future recommendations

Rossi, A., Hoogeveen, I. J., Bastek, V. B., de Boer, F., Montanari, C., Meyer, U., Maiorana, A., Bordugo, A., Dianin, A., Campana, C., Rigoldi, M., Kishnani, P. S., Pendyal, S., Strisciuglio, P., Gasperini, S., Parenti, G., Parini, R., Paci, S., Melis, D. & Derks, T. G. J., 1-Jul-2020, In: Journal of Inherited Metabolic Disease. 43, 4, p. 770-777 8 p.

Research priorities for liver glycogen storage disease: An international priority setting partnership with the James Lind Alliance

Peeks, F., Boonstra, W. F., de Baere, L., Carøe, C., Casswall, T., Cohen, D., Cowan, K., Ferrecchia, I., Ferriani, A., Gimbert, C., Landgren, M., Maldonado, N. L., McMillan, J., Nemeth, A., Seidita, N., Stachelhaus-Theimer, U., Weinstein, D. A. & Derks, T., Mar-2020, In: Journal of Inherited Metabolic Disease. 43, 2, p. 279-289 11 p.

Glycogen storage disease type 1a is associated with disturbed vitamin A metabolism and elevated serum retinol levels

Saeed, A., Hoogerland, J. A., Wessel, H., Heegsma, J., Derks, T. G. J., Veer, E., Mithieux, G., Rajas, F., Oosterveer, M. H. & Faber, K. N., 15-Jan-2020, In: Human Molecular Genetics. 29, 2, p. 264-273 10 p.

Efficacy and safety of D,L-3-hydroxybutyrate (D,L-3-HB) treatment in multiple acyl-CoA dehydrogenase deficiency

van Rijt, W. J., Jager, E. A., Allersma, D. P., Aktuğlu Zeybek, A. Ç., Bhattacharya, K., Debray, F-G., Ellaway, C. J., Gautschi, M., Geraghty, M. T., Gil-Ortega, D., Larson, A. A., Moore, F., Morava, E., Morris, A. A., Oishi, K., Schiff, M., Scholl-Bürgi, S., Tchan, M. C., Vockley, J., Witters, P. & 4 others, Wortmann, S. B., van Spronsen, F., Van Hove, J. L. K. & Derks, T. G. J., 6-Jan-2020, In: Genetics in Medicine. 22, 5, p. 908-916 9 p.

The need for additional care in patients with classical galactosaemia

Welling, L., Meester-Delver, A., Derks, T. G., Janssen, M. C. H., Hollak, C. E. M., de Vries, M. & Bosch, A. M., 23-Oct-2019, In: Disability and Rehabilitation. 41, 22, p. 2663-2668 6 p.

Transcriptome analysis suggests a compensatory role of the cofactors coenzyme A and NAD⁺ in medium-chain acyl-CoA dehydrogenase knockout mice

Martines, A-C. M. F., Gerding, A., Stolle, S., Vieira-Lara, M. A., Wolters, J. C., Jurdzinski, A., Bongiovanni, L., de Bruin, A., van der Vlies, P., van der Vries, G., Bloks, V. W., Derks, T. G. J., Reijngoud, D-J. & Bakker, B. M., 10-Oct-2019, In: Scientific Reports. 9, 1, 11 p., 14539.

A Nationwide Retrospective Observational Study Of Population Newborn Screening For Medium-Chain Acyl-CoA Dehydrogenase (MCAD) Deficiency In The Netherlands

Jager, E. A., Kuijpers, M. M., Bosch, A. M., Mulder, M. F., Rubio-Gozalbo, M. E., Visser, G., de Vries, M., Williams, M., Waterham, H. R., van Spronsen, F. J., Schielen, P. C. J. I. & Derks, T. G. J., Sep-2019, In: Journal of Inherited Metabolic Disease. 42, 5, p. 890-897 8 p.

Prediction of disease severity in multiple acyl-CoA dehydrogenase deficiency: a retrospective and laboratory cohort study

van Rijt, W. J., Ferdinandusse, S., Giannopoulos, P., Ruiter, J. P. N., de Boer, L., Bosch, A. M., Huidekoper, H. H., Rubio-Gozalbo, M. E., Visser, G., Williams, M., Wanders, R. J. A. & Derks, T. G. J., Sep-2019, In: Journal of Inherited Metabolic Disease. 42, 5, p. 878-889 12 p.

Changes in pediatric plasma acylcarnitines upon fasting for refined interpretation of metabolic stress

van Rijt, W. J., van der Ende, R. M., Volker-Touw, C. M. L., van Spronsen, F., Derks, T. G. J. & Heiner-Fokkema, M. R., Aug-2019, In: Molecular Genetics and Metabolism. 127, 4, p. 327-335 9 p.

Impact of NBS for VLCAD deficiency on genetic, enzymatic and clinical outcomes

Bleeker, J. C., Kok, I. L., Ferdinandusse, S., van der Pol, W. L., Cuppen, I., Bosch, A. M., Langeveld, M., Derks, T. G. J., Williams, M., de Vries, M., Mulder, M. F., Rubio Gozalbo, E., van der Velden, M. G. M. D. S., Rennings, A. J., Schielen, P. J. C. I., Dekkers, E., Houtkooper, R. H., Waterham, H. R., Pras-Raves, M. L., Wanders, R. J. A. & 4 others, van Hasselt, P. M., Schoenmakers, M., Wijburg, F. A. & Visser, G., May-2019, In: Journal of Inherited Metabolic Disease. 42, 3, p. 414-423 29 p.

Movement disorders and nonmotor neuropsychological symptoms in children and adults with classical galactosemia

Kuiper, A., Grünewald, S., Murphy, E., Coenen, M. A., Eggink, H., Zutt, R., Rubio-Gozalbo, M. E., Bosch, A. M., Williams, M., Derks, T. G. J., Lachmann, R. H. L., Brouwers, M. C. G. J., Janssen, M. C. H., Tijssen, M. A. & de Koning, T. J., May-2019, In: Journal of Inherited Metabolic Disease. 42, 3, p. 451-458 8 p.

The natural history of classic galactosemia: lessons from the GalNet registry

Rubio-Gozalbo, M. E., Haskovic, M., Bosch, A. M., Burnynte, B., Coelho, A. I., Cassiman, D., Couce, M. L., Dawson, C., Demirbas, D., Derks, T., Eyskens, F., Forga, M. T., Grunewald, S., Häberle, J., Hochuli, M., Hubert, A., Huidekoper, H. H., Janeiro, P., Kotzka, J., Knerr, I. & 23 others, Labrune, P., Landau, Y. E., Langendonk, J. G., Möslinger, D., Müller-Wieland, D., Murphy, E., Öunap, K., Ramadza, D., Rivera, I. A., Scholl-Buergi, S., Stepien, K. M., Thijs, A., Tran, C., Vara, R., Visser, G., Vos, R., de Vries, M., Waisbren, S. E., Welsink-Karssies, M. M., Wortmann, S. B., Gautschi, M., Treacy, E. P. & Berry, G. T., 27-Apr-2019, In: Orphanet journal of rare diseases. 14, 1, 11 p., 86.

Proposal for an individualized dietary strategy in patients with very long-chain acyl-CoA dehydrogenase deficiency

Bleeker, J. C., Kok, I. L., Ferdinandusse, S., de Vries, M., Derks, T. G. J., Mulder, M. F., Williams, M., Gozalbo, E. R., Bosch, A. M., van den Hurk, D. T., de Sain-van der Velden, M. G. M., Waterham, H. R., Wijburg, F. A. & Visser, G., Jan-2019, In: Journal of Inherited Metabolic Disease. 42, 1, p. 159-168 10 p.

A preliminary study of telemedicine for patients with hepatic glycogen storage disease and their healthcare providers: from bedside to home site monitoring

Hoogeveen, I. J., Peeks, F., de Boer, F., Lubout, C. M. A., de Koning, T. J., Te Boekhorst, S., Zandvoort, R.-J., Burghard, R., van Spronsen, F. J. & Derks, T. G. J., Dec-2018, In: Journal of Inherited Metabolic Disease. 41, 6, p. 929-936 8 p.

Next-generation glycogen storage diseases

Derks, T. G. J., Oosterveer, M. H. & De Souza, C. F., Dec-2018, In: Journal of Inherited Metabolic Disease. 41, 6, p. 911-912 2 p.

Pathogenic variants in glutamyl-tRNA(Gln) amidotransferase subunits cause a lethal mitochondrial cardiomyopathy disorder

Friederich, M. W., Timal, S., Powell, C. A., Dallabona, C., Kurolap, A., Palacios-Zambrano, S., Bratkovic, D., Derks, T. G. J., Bick, D., Bouman, K., Chatfield, K. C., Damouny-Naoum, N., Dishop, M. K., Falik-Zaccari, T. C., Fares, F., Fedida, A., Ferrero, I., Gallagher, R. C., Garesse, R., Gilberti, M. & 30 others, Gonzalez, C., Gowan, K., Habib, C., Halligan, R. K., Kalfon, L., Knight, K., Lefeber, D., Mamblona, L., Mandel, H., Mory, A., Ottoson, J., Paperna, T., Pruijn, G. J. M., Rebelo-Guioimar, P. F., Saada, A., Sainz, B., Salvemini, H., Schoots, M. H., Smeitink, J. A., Szukszto, M. J., ter Horst, H. J., van den Brandt, F., van Spronsen, F. J., Veltman, J. A., Wartchow, E., Wintjes, L. T., Zohar, Y., Fernandez-Moreno, M. A.,

Baris, H. N. & Donnini, C., 3-Oct-2018, In: Nature Communications. 9, 1, 14 p., 4065.

Safety issues associated with dietary management in patients with hepatic glycogen storage disease

Steunenberg, T. A. H., Peeks, F., Hoogeveen, I. J., Mitchell, J. J., Mundy, H., de Boer, F., Lubout, C. M. A., de Souza, C. F., Weinstein, D. A. & Derks, T. G. J., Sep-2018, In: Molecular Genetics and Metabolism. 125, 1-2, p. 79-85 7 p.

Glycogen storage disease type Ia adult presentation with microcytic anemia and liver adenomas

Moest, W., van der Deure, W., Koster, T., Spee-Dropková, M., Swart-Busscher, L., de Haas, R. J. & Derks, T. G. J., Aug-2018, In: Hepatology. 68, 2, p. 780-782 3 p.

Molybdenum cofactor deficiency type A: Prenatal monitoring using MRI

Lubout, C. M. A., Derks, T. G. J., Meiners, L., Erwich, J. J., Bergman, K. A., Luning, R. J., Schwarz, G., Veldman, A. & van Spronsen, F. J., May-2018, In: European Journal of Paediatric Neurology. 22, 3, p. 536-540 5 p.

Inborn Errors of Metabolism with Hypoglycemia Glycogen Storage Diseases and Inherited Disorders of Gluconeogenesis: Glycogen Storage Diseases and Inherited Disorders of Gluconeogenesis

Weinstein, D. A., Steuerwald, U., De Souza, C. F. M. & Derks, T. G. J., Apr-2018, In: Pediatric clinics of North America. 65, 2, p. 247-265 19 p.

Biallelic variants in WARS2 encoding mitochondrial tryptophanyl-tRNA synthase in six individuals with mitochondrial encephalopathy

Wortmann, S. B., Timal, S., Venselaar, H., Wintjes, L. T., Kopajtich, R., Feichtinger, R. G., Onnekink, C., Muhlmeister, M., Brandt, U., Smeitink, J. A., Veltman, J. A., Sperl, W., Lefeber, D., Puijn, G., Stojanovic, V., Freisinger, P., von Spronsen, F., Derks, T. G. J., Veenstra-Knol, H. E., Mayr, J. A. & 4 others, Rotig, A., Tarnopolsky, M., Prokisch, H. & Rodenburg, R. J., Dec-2017, In: Human Mutation. 38, 12, p. 1786-1795 10 p.

Hepatocytes Contribute to Residual Glucose Production in a Mouse Model for Glycogen Storage Disease Type Ia

Hijmans, B. S., Boss, A., van Dijk, T. H., Soty, M., Wolters, H., Mutel, E., Groen, A. K., Derks, T. G. J., Mithieux, G., Heerschap, A., Reijngoud, D.-J., Rajas, F. & Oosterveer, M. H., Dec-2017, In: Hepatology. 66, 6, p. 2042-2054 13 p.

Clinical and biochemical heterogeneity between patients with glycogen storage disease type IA: the added value of CUSUM for metabolic control

Peeks, F., Steunenberg, T. A. H., de Boer, F., Rubio-Gozalbo, M. E., Williams, M., Burghard, R., Rajas, F., Oosterveer, M. H., Weinstein, D. A. & Derks, T. G. J., Sep-2017, In: Journal of Inherited Metabolic Disease. 40, 5, p. 695-702 8 p.

Clinical presentation and outcome in a series of 32 patients with 2-methylacetoacetyl-coenzyme A thiolase (MAT) deficiency

Grünert, S. C., Schmitt, R. N., Schlatter, S. M., Gemperle-Britschgi, C., Balci, M. C., Berg, V., Çoker, M., Das, A. M., Demirkol, M., Derks, T. G. J., Gökçay, G., Uçar, S. K., Konstantopoulou, V., Christoph Korenke, G., Lotz-Havla, A. S., Schlune, A., Staufner, C., Tran, C., Visser, G., Schwab, K. O. & 2 others, Fukao, T. & Sass, J. O., Sep-2017, In: Molecular Genetics and Metabolism. 122, 1-2, p. 67-75 9 p.

Nine years of newborn screening for classical galactosemia in the Netherlands: Effectiveness of screening methods, and identification of patients with previously unreported phenotypes

Welling, L., Boelen, A., Derks, T. G. J., Schielen, P. C. J. I., Vries, M. D., Williams, M., Wijburg, F. A. & Bosch, A. M., Mar-2017, In: Molecular Genetics and Metabolism. 120, 3, p. 223-228 6 p.

Inflammatory Bowel Disease in Glycogen Storage Disease Type Ia: A Case Series

Lawrence, N. T., Chengsupanimit, T., Brown, L. M., Derks, T. G. J., Smit, G. P. A. & Weinstein, D. A., Feb-2017, In: Journal of Pediatric Gastroenterology and Nutrition. 64, 2, p. 52-54 3 p.

What Is the Best Blood Sampling Time for Metabolic Control of Phenylalanine and Tyrosine Concentrations in Tyrosinemia Type 1 Patients?

van Dam, E., Daly, A., Venema-Liefaard, G., van Rijn, M., Derks, T. G. J., McKiernan, P. J., Heiner-Fokkema, R., MacDonald, A. & van Spronsen, F. J., 25-Jan-2017, *JIMD Reports*. Morava, E., Baumgartner, M., Patterson, M., Rahman, S., Zschocke, J. & Peters, V. (eds.). Springer, p. 49-57 9 p. (Journal of Inherited Metabolic Disorders; vol. 36).

Living on the edge: substrate competition explains loss of robustness in mitochondrial fatty-acid oxidation disorders
van Eunen, K., Volker-Touw, C. M. L., Gerding, A., Bleeker, A., Wolters, J. C., Rijt, van, W., Martines, A-C. M. F., Niezen-Koning, K. E., Heiner, R. M., Permentier, H., Groen, A. K., Reijngoud, D-J., Derks, T. G. J. & Bakker, B. M., 7-Dec-2016, In: BMC Biology. 14, 1, 15 p., 107.

Neonates at risk of medium-chain acyl-CoA dehydrogenase deficiency: a perinatal protocol for use before population neonatal screening test results become available
van Rijt, W. J., Jager, E. A., Spronsen, van, F. J., de Koning, T., Heiner-Fokkema, M. R. & Derks, T. G. J., Dec-2016, In: Genetics in Medicine. 18, 12, p. 1322-1323 2 p.

Glycogen storage disease type III: diagnosis, genotype, management, clinical course and outcome
Sentner, C. P., Hoogeveen, I. J., Weinstein, D. A., Santer, R., Murphy, E., McKiernan, P. J., Steuerwald, U., Beauchamp, N. J., Taybert, J., Laforet, P., Petit, F. M., Hubert, A., Labrune, P., Smit, G. P. A. & Derks, T. G. J., Sep-2016, In: Journal of Inherited Metabolic Disease. 39, 5, p. 697-704 8 p.

Childhood Pompe disease: clinical spectrum and genotype in 31 patients
van Capelle, C. I., van der Meijden, J. C., van den Hout, J. M. P., Jaeken, J., Baethmann, M., Voit, T., Kroos, M. A., Derks, T. G. J., Rubio-Gozalbo, M. E., Willemsen, M. A., Lachmann, R. H., Mengel, E., Michelakakis, H., de Jongste, J. C., Reuser, A. J. J. & van der Ploeg, A. T., 18-May-2016, In: Orphanet journal of rare diseases. 11, 1, 11 p., 65.

Analysis of body composition and nutritional status in Brazilian phenylketonuria patients
Mazzola, P. N., Nalin, T., Castro, K., van Rijn, M., Derks, T. G. J., Perry, I. D. S., Mainieri, A. S. & Schwartz, I. V. D., Mar-2016, In: Molecular genetics and metabolism reports. 6, p. 16-20 5 p.

Orthotopic liver transplantation in glycogen storage disease type Ia: Perioperative glucose and lactate homeostasis
Wilke, M. V. M. B., De Kleine, R. H., Wietasch, J. K. G., Van Amerongen, C. C. A., Blokzijl, H., Van Spronsen, F. J., Schwartz, I. V. D. & Derks, T. G. J., 25-Feb-2016, In: Journal of Inborn Errors of Metabolism and Screening. 4

Muscle Ultrasound in Patients with Glycogen Storage Disease Types I and III
Verbeek, R. J., Sentner, C. P., Smit, G. P. A., Maurits, N. M., Derks, T. G. J., van der Hoeven, J. H. & Sival, D. A., Jan-2016, In: Ultrasound in Medicine and Biology. 42, 1, p. 133-142 10 p.

Inborn Errors of Metabolism That Cause Sudden Infant Death: A Systematic Review with Implications for Population Neonatal Screening Programmes
van Rijt, W. J., Koolhaas, G. D., Bekhof, J., Fokkema, M. R. H., de Koning, T. J., Visser, G., Schielen, P. C. J. I., Spronsen, van, F. & Derks, T. G. J., 2016, In: Neonatology. 109, 4, p. 297-302 6 p.

Normoglycemic Ketonemia as Biochemical Presentation in Ketotic Glycogen Storage Disease
Hoogeveen, I. J., van der Ende, R. M., van Spronsen, F. J., de Boer, F., Heiner-Fokkema, R. & Derks, T. G. J., 2016, *JIMD Reports*. Morava, E., Baumgartner, M., Patterson, M., Rahman, S., Zschocke, J. & Peters, V. (eds.). Springer, Vol. 28. p. 41-47 7 p. (Journal of Inherited Metabolic Disorders; vol. 28).

Acute exercise in treated phenylketonuria patients: Physical activity and biochemical response
Mazzola, P. N., Teixeira, B. C., Schirmbeck, G. H., Reischak-Oliveira, A., Derks, T. G. J., van Spronsen, F. J., Dutra-Filho, C. S. & Schwartz, I. V. D., Dec-2015, In: Molecular genetics and metabolism reports. 5, p. 55-59 5 p.

Efficacy and safety of cyclic pyranopterin monophosphate substitution in severe molybdenum cofactor deficiency type A: a prospective cohort study
Schwahn, B. C., Van Spronsen, F. J., Belaidi, A. A., Bowhay, S., Christodoulou, J., Derks, T. G., Hennermann, J. B., Jameson, E., Koenig, K., McGregor, T. L., Font-Montgomery, E., Santamaria-Araujo, J. A., Santra, S., Vaidya, M., Vierzig, A., Wassmer, E., Weis, I., Wong, F. Y., Veldman, A. & Schwarz, G., 14-Nov-2015, In: The Lancet. 386, 10007, p. 1955-1963 9 p.

Determination of amylose/amylopectin ratio of starches

Nalin, T., Sperb-Ludwig, F., Venema, K., Derks, T. G. J. & Schwartz, I. V. D., Sep-2015, In: Journal of Inherited Metabolic Disease. 38, 5, p. 985-986 2 p.

Erratum to: In vitro digestion of starches in a dynamic gastrointestinal model: an innovative study to optimize dietary management of patients with hepatic glycogen storage diseases (vol 38, pg 529, 2015)

Nalin, T., Venema, K., Weinstein, D. A., de Souza, C. F. M., Perry, I. D. S., van Wandelen, M. T. R., van Rijn, M., Smit, G. P. A., Schwartz, I. V. D. & Derks, T. G. J., Sep-2015, In: Journal of Inherited Metabolic Disease. 38, 5, p. 987 1 p.

Inborn errors in fatty-acid metabolism: Living on the edge

van Eunen, K., Touw, C. M. L., Gerding, A., Bleeker, A., Wolters, K., Heiner, R., Derks, T. G. J., Permentier, H., Reijngoud, D. -J., Groen, A. K. & Bakker, B. M., Jul-2015, In: Febs Journal. 282, Supplement S1, p. 24 1 p., P28-003-SH.

Dietary management in glycogen storage disease type III: What is the evidence?

Derks, T. G. J. & Smit, G. P. A., May-2015, In: Journal of Inherited Metabolic Disease. 38, 3, p. 545-550 6 p.

Evaluation of glycogen storage disease as a cause of ketotic hypoglycemia in children

Brown, L. M., Corrado, M. M., van der Ende, R. M., Derks, T. G. J., Chen, M. A., Siegel, S., Hoyt, K., Correia, C. E., Lumpkin, C., Flanagan, T. B., Carreras, C. T. & Weinstein, D. A., May-2015, In: Journal of Inherited Metabolic Disease. 38, 3, p. 489-493 5 p.

In vitro digestion of starches in a dynamic gastrointestinal model: an innovative study to optimize dietary management of patients with hepatic glycogen storage diseases

Nalin, T., Venema, K., Weinstein, D. A., de Souza, C. F. M., Perry, I. D. S., van Wandelen, M. T. R., van Rijn, M., Smit, G. P. A., Schwartz, I. V. D. & Derks, T. G. J., May-2015, In: Journal of Inherited Metabolic Disease. 38, 3, p. 529-536 8 p.

Lipids in hepatic glycogen storage diseases: Pathophysiology, monitoring of dietary management and future directions

Derks, T. G. J. & van Rijn, M., May-2015, In: Journal of Inherited Metabolic Disease. 38, 3, p. 537-543 7 p.

BH4 treatment in BH4-responsive PKU patients: Preliminary data on blood prolactin concentrations suggest increased cerebral dopamine concentrations

van Vliet, D., Anjema, K., Jahja, R., de Groot, M. J., Liemburg, G. B., Heiner-Fokkema, R., van der Zee, E. A., Derks, T. G. J., Kema, I. P. & van Spronsen, F. J., Jan-2015, In: Molecular Genetics and Metabolism. 114, 1, p. 29-33 5 p.

Infants with Tyrosinemia Type 1: Should phenylalanine be supplemented?

van Vliet, D., van Dam, E., van Rijn, M., Derks, T. G. J., Venema-Liefaard, G., Hitzert, M. M., Lunsing, R. J., Heiner-Fokkema, M. R. & van Spronsen, F. J., 2015, *JIMD Reports*. Zschocke, J., Baumgartner, M., Morava, E., Patterson, M. & Rahman, S. (eds.). Springer, p. 117-124 8 p. (Journal of Inherited Metabolic Disorders; vol. 18).

Favorable outcome after physiologic dose of sodium-D,L-3-hydroxybutyrate in severe MADD

Van Rijn, W. J., Heiner-Fokkema, M. R., Sarvaas, G. J. D. M., Waterham, H. R., Blokpoel, R. G. T., van Spronsen, F. J. & Derks, T. G. J., Oct-2014, In: Pediatrics. 134, 4, p. e1224-e1228 5 p.

From genome to phenome-Simple inborn errors of metabolism as complex traits

Touw, C. M. L., Derks, T. G. J., Bakker, B. M., Groen, A. K., Smit, G. P. A. & Reijngoud, D. J., Oct-2014, In: Biochimica et biophysica acta. 1842, 10, p. 2021-2029 9 p.

Experimental evidence for protein oxidative damage and altered antioxidant defense in patients with medium-chain acyl-CoA dehydrogenase deficiency

Derks, T. G. J., Touw, C. M. L., Ribas, G. S., Biancini, G. B., Vanzin, C. S., Negretto, G., Mescka, C. P., Reijngoud, D. J., Smit, G. P. A., Wajner, M. & Vargas, C. R., Sep-2014, In: Journal of Inherited Metabolic Disease. 37, 5, p. 783-789 7 p.

Recombinant phenylalanine ammonia lyase in phenylketonuria

van Spronsen, F. J. & Derks, T. G. J., 5-Jul-2014, In: The Lancet. 384, 9937, p. 6-8 3 p.

Mutations in RARS cause hypomyelination

Wolf, N. I., Salomons, G. S., Rodenburg, R. J., Pouwels, P. J. W., Schieving, J. H., Derks, T. G. J., Fock, J. M., Rump, P., van Beek, D. M., van der Knaap, M. S. & Waisfisz, Q., Jul-2014, In: *Annals of Neurology*. 76, 1, p. 134-139 6 p.

Single amino acid supplementation in aminoacidopathies: a systematic review

van Vliet, D., Derks, T. G. J., van Rijn, M., de Groot, M. J., MacDonald, A., Heiner-Fokkema, M. R. & van Spronsen, F. J., 13-Jan-2014, In: *Orphanet journal of rare diseases*. 9, 14 p., 7.

Dietary treatment of glycogen storage disease type Ia: Uncooked cornstarch and/or continuous nocturnal gastric drip-feeding?

Derks, T. G. J., Martens, D. H., Sentner, C. P., van Rijn, M., de Boer, F., Smit, G. P. A. & van Spronsen, F. J., May-2013, In: *Molecular Genetics and Metabolism*. 109, 1, p. 1-2 2 p.

In vitro and in vivo consequences of variant medium-chain acyl-CoA dehydrogenase genotypes

Touw, C. M. L., Smit, G. P. A., Niezen-Koning, K. E., Bosgraaf-de Boer, C., Gerding, A., Reijngoud, D.-J. & Derks, T., 20-Mar-2013, In: *Orphanet journal of rare diseases*. 8, 8 p., 43.

Risk stratification by residual enzyme activity after newborn screening for medium-chain acyl-CoA dehydrogenase deficiency: data from a cohort study

Touw, C. M. L., Smit, G. P. A., de Vries, M., de Klerk, J. B. C., Bosch, A. M., Visser, G., Mulder, M. F., Rubio-Gozalbo, M. E., Elvers, B., Niezen-Koning, K. E., Wanders, R. J. A., Waterham, H. R., Reijngoud, D.-J. & Derks, T. G. J., 25-May-2012, In: *Orphanet journal of rare diseases*. 7, 8 p., 30.

Sweet and sour aspects of medium-chain acyl CoA dehydrogenase deficiency. Commentary on K. Yusuf et al. Neonatal ventricular tachyarrhythmias in medium chain acyl-CoA dehydrogenase deficiency (*Neonatology* 2010;98:260-264)

Derks, T. G. J., 2010, In: *Neonatology*. 98, 3, p. 265-267 3 p.

Disturbed hepatic carbohydrate management during high metabolic demand in medium-chain acyl-CoA dehydrogenase (MCAD)-deficient mice

Herrema, H., Derks, T., van Dijk, T. H., Bloks, V. W., Gerding, A., Havinga, R., Tietge, U. J. F., Müller, M., Smit, G. P. A., Kuipers, F. & Reijngoud, D.-J., Jun-2008, In: *Hepatology*. 47, 6, p. 1894-1904 11 p.

Inhibition of mitochondrial fatty acid oxidation in vivo only slightly suppresses gluconeogenesis but enhances clearance of glucose in mice

Derks, T. G. J., van Dijk, T. H., Grefhorst, A., Rake, J.-P., Smit, G. P. A., Kuipers, F. & Reijngoud, D.-J., Mar-2008, In: *Hepatology*. 47, 3, p. 1032-1042 11 p.

Neonatal screening for medium-chain acyl-CoA dehydrogenase (MCAD) deficiency in The Netherlands: The importance of enzyme analysis to ascertain true MCAD deficiency

Derks, T., Boer, T. S., van Assen, A., Bos, T., Ruiter, J. H., Waterham, H. R., Niezen-Koning, K. E., Wanders, R. J. A., Rondeel, J. M. M., Loeber, J. G., ten Kate, L. P., Smit, G. P. A. & Reijngoud, D. -J., Feb-2008, In: *Journal of Inherited Metabolic Disease*. 31, 1, p. 88-96 9 p.

Cost-effectiveness of neonatal screening for medium chain acyl-CoA dehydrogenase deficiency: The homogeneous population of the Netherlands

van der Hilst, C. S., Derks, T. G. J., Reijngoud, D.-J., Smit, G. P. A. & ten Vergert, E., Aug-2007, In: *Journal of Pediatrics*. 151, 2, p. 115-120 6 p.

Safe and unsafe duration of fasting for children with MCAD deficiency

Derks, T. G. J., van Spronsen, F. J., Rake, J. P., van der Hilst, C. S., Span, M. M. & Smit, G. P. A., Jan-2007, In: *European Journal of Pediatrics*. 166, 1, p. 5-11 7 p.

MCAD deficiency: clinical and laboratory studies

Derks, T. G. J., 2007, s.n.. 163 p.

The natural history of medium-chain acyl CoA dehydrogenase deficiency in the Netherlands: Clinical presentation and outcome

Derks, T. G. J., Reijngoud, D.-J., Waterham, H. R., Gerver, W.-J. M., van den Berg, M. P., Sauer, P. J. J. & Smit, G. P. A., May-2006, In: Journal of Pediatrics. 148, 5, p. 665-670 6 p.

Acute hepatic steatosis in mice by blocking beta-oxidation does not reduce insulin sensitivity of very-low-density lipoprotein production

Grefhorst, A., Hoekstra, J., Derks, T. G. J., Ouwens, D. M., Baller, J. F. W., Havinga, R., Havekes, L. M., Romijn, J. A. & Kuipers, F., Sep-2005, In: American Journal of Physiology-Gastrointestinal and Liver Physiology. 289, 3, p. G592-G598 7 p.

The difference between observed and expected prevalence of MCAD deficiency in The Netherlands: a genetic epidemiological study

Derks, T. G. J., Duran, M., Waterham, H. R., Reijngoud, D.-J., Ten Kate, L. P. & Smit, G. P. A., Aug-2005, In: European Journal of Human Genetics. 13, 8, p. 947-952 6 p.

Deficiëntie van het vetzuuroxidatie-enzym middenketen-acyl-coënzym-A-dehydrogenase (MCAD) bij een volwassene, opgespoord tijdens een proefproject voor neonatale screening]

Derks, T. G., Jakobs, H., Gerding, A., Niezen-Koning, K. E., Reijngoud, D. J. & Smit, G. P. A., 30-Oct-2004, In: Nederlands Tijdschrift voor de Geneeskunde. 148, 44, p. 2185-2190 6 p.

Activities

International GSD Conference

Terry Derks (Organiser)
15-Jun-2017 → 17-Jun-2017

European Metabolic Group (EMG) - Zagreb, Croatia - May 2017

Terry Derks (Organiser)
25-May-2017 → 27-May-2017

Patient organization VKS (Netherlands) - MADD patients' meeting 2017

Terry Derks (Keynote speaker)
28-Jan-2017

Encontro medicos e pacientes de Glicogenose Brasil - Ribeirao Preto, Brazil - Nov 2016

Terry Derks (Keynote speaker)
26-Nov-2016 → 27-Nov-2016

Patient organization VKS (Netherlands) - GSD patients' meeting 2016

Terry Derks (Keynote speaker)
28-May-2016

Scandinavian Association for Glycogen Storage Disease - 3rd Conference - Ängelholm, Sweden - May 2016

Terry Derks (Keynote speaker)
14-May-2016 → 15-May-2016

ESN Voorjaarssymposium

Terry Derks (Organiser)
19-Apr-2016 → 20-Apr-2016

ESN nascholing 2016

Terry Derks (Speaker)
2016

XI Course on Diagnosis and Treatment of Metabolic Disorders

Terry Derks (Attendee)
29-Nov-2015 → 4-Dec-2015

Medische Publieksacademie UMCG

Terry Derks (Keynote speaker)
12-Oct-2015

GSMSPHD conference

Terry Derks (Invited speaker)
12-Jun-2015

Patient organization VKS (Netherlands) - MCADD patients' meeting 2015

Terry Derks (Keynote speaker)
30-May-2015

Jahresfachtagung „SHG Glykogenose Deutschland e.V.“ - Duderstadt, Germany - March 2015

Terry Derks (Invited speaker)
27-Mar-2015 → 29-Mar-2015

International Network for Fatty Acid Oxidation Research and Management

Terry Derks (Invited speaker)
2015

Society for the Study of Inborn Errors of Metabolism (SSIEM)

Terry Derks (Keynote speaker)
2015 → ...

Recordati-Rare-Diseases-Academy Symposium on Changing Spectrum of IMD - Surviving Longer and Growing Old with IMDs

Terry Derks (Keynote speaker)
2014 → ...

International GSD Conference

Terry Derks (Keynote speaker)
2013 → ...

Journal of Inborn Errors of Metabolism and Screening (Journal)

Terry Derks (Editorial board member)
2013 → ...

Medical Sciences Summer School Pediatrics

Terry Derks (Organiser)
2013 → ...

Press clippings**JIMD Podcast**

Terry Derks
28/05/2021
1 Media contribution

GSD 1b - Dr Terry Derks - A Podcast on GSD, symptoms, treatments & what daily life looks like

Terry Derks
13/08/2020
2 Media contributions

Fasting Tolerance in MCADD-infants (FITtING MCADD)

Terry Derks
03/12/2018
1 Media contribution

ClinicalTrials.gov Identifier: NCT03517085

Terry Derks
13/03/2019
1 Media contribution

ClinicalTrials.gov Identifier: NCT02318966

Terry Derks
11/10/2018
1 Media contribution

ClinicalTrials.gov Identifier: NCT03011203

Terry Derks
01/05/2018
1 Media contribution

ClinicalTrials.gov Identifier: NCT03761693

Terry Derks
03/12/2018
1 Media contribution

International Liver Glycogen Storage Disease (IGSD) Priority Setting Partnership

Terry Derks
16/06/2017
1 Media contribution

Lezing Publieksacademie

Terry Derks
12/10/2015
1 Media contribution

Member of the UEF-JSM Talent Grant Committee

Terry Derks
01/01/2012
1 item of Media coverage

Clinical trial: Glycosade v UCCS in the Dietary Management of Hepatic GSD (Glyde) (ClinicalTrials.gov Identifier: NCT02318966)

Terry Derks & Irene Hoogeveen
03/02/2017
1 item of Media coverage

Clinical trial: Acute Nutritional Ketosis in GSD IIIa (ClinicalTrials.gov Identifier: NCT03011203)

Terry Derks, Irene Hoogeveen & Jeroen Jeneson
22/12/2016
1 item of Media coverage

Retrospective international study on Betahydroxybutyrate (3-HB) in multiple acyl CoA dehydrogenase deficiency

Terry Derks & Willemijn Rijt, van

06/02/2017

1 item of Media coverage

Test filtert ziek kind eruit

Terry Derks

10/10/2015

1 item of Media coverage

Kenniskaart vetzuuroxidatie stoornissen

Terry Derks

04/05/2015

1 item of Media coverage

Kenniskaart GSD

Terry Derks

06/05/2016

1 item of Media coverage

YouTube movie: "De weg naar een diagnose"

Terry Derks

08/07/2015

1 item of Media coverage

YouTube movie: "Diagnose: Achter de schermen"

Terry Derks

08/07/2015

1 item of Media coverage

Press clippings

JIMD Podcast

Terry Derks

28/05/2021

1 Media contribution

GSD 1b - Dr Terry Derks - A Podcast on GSD, symptoms, treatments & what daily life looks like

Terry Derks

13/08/2020

2 Media contributions

ClinicalTrials.gov Identifier: NCT03517085

Terry Derks

13/03/2019

1 Media contribution

ClinicalTrials.gov Identifier: NCT03761693

Terry Derks

03/12/2018

1 Media contribution

Fasting Tolerance in MCADD-infants (FiTiNg MCADD)

Terry Derks

03/12/2018

1 Media contribution

ClinicalTrials.gov Identifier: NCT02318966

Terry Derks
11/10/2018
1 Media contribution

ClinicalTrials.gov Identifier: NCT03011203

Terry Derks
01/05/2018
1 Media contribution

International Liver Glycogen Storage Disease (IGSD) Priority Setting Partnership

Terry Derks
16/06/2017
1 Media contribution

Retrospective international study on Betahydroxybutyrate (3-HB) in multiple acyl CoA dehydrogenase deficiency

Terry Derks & Willemijn Rijt, van
06/02/2017
1 item of Media coverage

Clinical trial: Glycosade v UCCS in the Dietary Management of Hepatic GSD (Glyde) (ClinicalTrials.gov Identifier: NCT02318966)

Terry Derks & Irene Hoogeveen
03/02/2017
1 item of Media coverage

Clinical trial: Acute Nutritional Ketosis in GSD IIIa (ClinicalTrials.gov Identifier: NCT03011203)

Terry Derks, Irene Hoogeveen & Jeroen Jeneson
22/12/2016
1 item of Media coverage

Kenniskaart GSD

Terry Derks
06/05/2016
1 item of Media coverage

Lezing Publieksacademie

Terry Derks
12/10/2015
1 Media contribution

Test filtert ziek kind eruit

Terry Derks
10/10/2015
1 item of Media coverage

YouTube movie: "De weg naar een diagnose"

Terry Derks
08/07/2015
1 item of Media coverage

YouTube movie: "Diagnose: Achter de schermen"

Terry Derks
08/07/2015
1 item of Media coverage

Kenniskaart vetzuuroxidatie stoornissen

Terry Derks

04/05/2015

1 item of Media coverage

Member of the UEF-JSM Talent Grant Committee

Terry Derks

01/01/2012

1 item of Media coverage