# Publications Univ.-Prof. Dr. med. Joachim Weis

## Contents



## 20 selected publications

**Weis J**, Schröder JM: Differential effects of nerve, muscle and fat tissue on regenerating nerve fibers in vivo. *Muscle & Nerve* 12: 723-734, 1989

**Weis J**, Fine SM, David C, Savarirayan S, Sanes JR: Integration site-dependent expression of a transgene reveals special features of cells associated with neuromuscular junctions. *J Cell Biol* 113: 1385-1397, 1991

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Porter BE, **Weis J**, Merlie JP, Sanes JR: A motoneuron-selective stop signal for neurite outgrowth in the synaptic protein, s-laminin. *Neuron* 14: 549-559, 1995

**Weis J**, Lie DC, Ragoß U, Züchner S, Karpati G, Yancopoulos GD, DiStefano PS: CNTF receptor alpha expression is increased in acute denervation atrophy of human skeletal muscle. *J Neuropathol Exp Neurol* 57: 850-857, 1998

Brunello AG, Weissenberger J, Kappeler A, Vallan C, Peters M, Rose-John S, **Weis J**: CNS alterations in interleukin-6/soluble interleukin-6 receptor alpha double transgenic mice. *Am J Pathol* 157: 1485-1493, 2000

Weissenberger J, Loeffler S, Afanasieva T, Aguzzi A, Kopf M, **Weis J**: IL-6 is required for glioma development in a mouse model. *Oncogene* 23: 3308-3316, 2004

Bremer J, Baumann F, Tiberi C, Wessig C, Fischer H, Schwarz P, Steele AD, Toyka KV, Nave KA, **Weis J**, Aguzzi A. Axonal prion protein is required for peripheral myelin maintenance. *Nature Neurosci.* 13(3): 310-318, 2010

Brauers E, Dreier A, Roos A, Wormland B, **Weis J***, Krüttgen A*. Differential effects of myopathy-associated caveolin 3-mutants on growth factor signaling. *Am J Pathol.* 177(1): 261-70, 2010 *Equal contribution

**Weis J***, Katona I*, Müller-Newen G, Sommer C, Necula G, Hendrich C, Ludolph AC, Sperfeld A-D. Small fiber neuropathy in ALS patients. *Neurology.* 76(23): 2024-2029, 2011. *Equal contribution

Prause J*, Goswami A*, Katona I, Roos A, Schnizler M, Bushuven E, Dreier A, Buchkremer S, Johann S, Beyer C, Deschauer M, Troost D, **Weis J**. Altered localization, abnormal modification and loss of function of Sigma receptor-1 in amyotrophic lateral sclerosis. *Hum Mol Genet.* 22(8): 1581-1600, 2013. *Equal contr.

Roos A, Buchkremer S, Kollipara L, Labisch T, Gatz C, Zitzelsberger M, Brauers E, Nolte K, Schröder JM, Kirschner J, Jesse CM, Goebel HH, Goswami A, Zimmermann R, Zahedi RP, Senderek J, **Weis J**. Myopathy in Marinesco-Sjögren syndrome links endoplasmic reticulum chaperone dysfunction to nuclear envelope pathology. *Acta Neuropathol*. 127(5): 761-777, 2014

Filézac de L'Etang A, Maharjan N, Cordeiro Braña M, Ruegsegger C, Rehmann R, Goswami A, Roos A, Troost D, Schneider BL, **Weis J**, Saxena S. Marinesco-Sjögren syndrome protein SIL1 regulates motor neuron subtype-selective ER stress in ALS. *Nature Neurosci.* 18(2): 227-238, 2015

Khaminets A, Heinrich T, Mari M, Grumati P, Huebner AK, Akutsu M, Liebmann L, Stolz A, Nietzsche S, Koch N, Mauthe M, Katona I, Qualmann B, **Weis J**, Reggiori F, Kurth I, Hübner CA, Dikic I. Regulation of endoplasmic reticulum turnover by selective autophagy. *Nature.* 522 (7556): 354-358, 2015

Goswami A, Jesse C, Chandrasekar A, Bushuven E, Vollrath J, Dreser A, Katona I, Beyer C, Johann S, Feller A, Grond M, Wagner S, Nikolin S, Troost D, **Weis J**. Accumulation of STIM1 is associated with the degenerative muscle fibre phenotype in ALS and other neurogenic atrophies. *Neuropathol Appl Neurobiol*. 41(3): 304-18, 2015

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Bouhy D, Juneja M, Katona I, Holmgren A, Asselbergh B, De Winter V, Hochepied T, Goossens S, Haigh JJ, Libert C, Ceuterick-de Groote C, Irobi J, **Weis J**, Timmerman V. A knock-in/knock-out mouse model of HSPB8-associated distal hereditary motor neuropathy and myopathy reveals toxic gain-of-function of mutant Hspb8. *Acta Neuropathol.* 135(1): 131-148, 2018

Mair D, Biskup S, Kress W, Abicht A, Brück W, Zechel S, Knop KC, Koenig FB, Tey S, Nikolin S, Eggermann K, Kurth I, Ferbert A, **Weis J**. Differential diagnosis of vacuolar myopathies in the NGS era. *Brain Pathol.* 30(5): 877-896, 2020

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**Chronological list of publications in scientific journals excluding reviews, guidelines and comments**

**Publications 2022**

364. von Stillfried S, Bülow RD, Röhrig R, Boor P; German Registry of COVID-19 Autopsies (DeRegCOVID), DeRegCOVID Collaborators. First report from the German COVID-19 autopsy registry. *Lancet Reg Health Eur.* 2022 Feb 18; 15: 100330. doi: 10.1016/j.lanepe.2022.100330. eCollection 2022 Apr. PMID: 35531493

363. von Stillfried S, Bülow RD, Röhrig R, Meybohm P, Boor P; German Registry of COVID-19 Autopsies (DeRegCOVID), DeRegCOVID Collaborators#. Intracranial hemorrhage in COVID-19 patients during extracorporeal membrane oxygenation for acute respiratory failure: a nationwide register study report. *Crit Care.* 2022 Mar 28; 26(1): 83. doi: 10.1186/s13054-022-03945-x. PMID: 35346314

362. Pilotto F, Schmitz A, Maharjan N, Diab R, Odriozola A, Tripathi P, Yamoah A, Scheidegger O, Oestmann A, Dennys CN, Sinha Ray S, Rodrigo R, Kolb S, Aronica E, Di Santo S, Widmer HR, Charlet-Berguerand N, Selvaraj BT, Chandran S, Meyer K, Zuber B, Goswami A, **Weis J**, Saxena S. PolyGA targets the ER stress-adaptive response by impairing GRP75 function at the MAM in *C9ORF72*-ALS/FTD. *Acta Neuropathol 2*022 144(5):939-966, 2022

361. Maier A, Kapfenberger R, Katona I, **Weis J**, Schulz JB, Rolke R. Nonregional small fibre neuropathy in cases of autoimmune autonomic neuropathy. *J Neurol.* 2022 Sep 9. Epub ahead of print.

360. Altinova H, Achenbach P, Palm M, Katona I, Hermans E, Clusmann H, **Weis J**, Brook GA. Characterization of a Novel Aspect of Tissue Scarring Following Experimental Spinal Cord Injury and the Implantation of Bioengineered Type-I Collagen Scaffolds in the Adult Rat: Involvement of Perineurial-Like Cells? *Int. J. Mol. Sci.* 23(6), 3221, 2022

359. Dohrn MF, Heller C, Zengeler D, Obermaier CD, Biskup S, **Weis J**, Nikolin S, Claeys KG, Schöne U, Beijer D, Winter N, Achenbach P, Gess B, Schulz JB, Mulahasanovic L. Heterozygous POLG variant Ser1181Asn co-segregating in a family with autosomal dominant axonal neuropathy, proximal muscle fatigability, ptosis, and ragged red fibers. *Neurol Res Pract.* 4(1): 5, 2022

358. Koeppen S, Hense J, Nolte KW, **Weis J**. Immune-mediated neuropathy related to bortezomib in a patient with multiple myeloma. *Arch Pathol Clin Res*. 6: 001-004, 2022

**Publications 2021**

357. Ferreira N, Richner M, van der Laan A, Bergholdt Jul Christiansen I, Vægter CB, Nyengaard JR, Halliday GM, **Weis J**, Giasson BI, Mackenzie IR, Jensen PH, Jan A. Prodromal neuroinvasion of pathological α-synuclein in brainstem reticular nuclei and white matter lesions in a model of α-synucleinopathy. *Brain Commun.* 3(2): fcab104, 2021

356. Della Marina A, Arlt A, Schara-Schmidt U, Depienne C, Gangfuß A, Kölbel H,Sickmann A, Freier E, Kohlschmidt N, Hentschel A, **Weis J**, Czech A, Grüneboom A,

Roos A. Phenotypical and Myopathological Consequences of Compound Heterozygous Missense and Nonsense Variants in SLC18A3. *Cells*. 10(12):3481, 2021

355. Hummel C, Leylamian O, Pösch A, **Weis J**, Aronica E, Beyer C, Johann S. Expression and cell type-specific localization of inflammasome sensors in the spinal cord of SOD1(G93A) mice and sALS patients. *Neuroscience.* 463: 288-302, 2021

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348. Deschauer M, Hengel H, Rupprich K, Kreiß M, Schlotter-Weigel B, Grimmel M, Admard J, Schneider I, Alhaddad B, Gazou A, Sturm M, Vorgerd M, Balousha G, Balousha O, Falna M, Kirschke JS, Kornblum C, Jordan B, Kraya T, Strom TM, **Weis J**, Schöls L, Schara U, Zierz S, Riess O, Meitinger T, Haack TB. Bi-allelic truncating mutations in VWA1 cause neuromyopathy. *Brain* 144(2):574-583, 2021

347. Lausberg E, Gießelmann S, Dewulf JP, Wiame E, Holz A, Salvarinova R, Van Karnebeek C, Klemm P, Ohl K, Mull M, Braunschweig T, **Weis J**, Sommer C, Demuth S, Haase C, Debray F-G, Libioulle C, Choukair D, Oommen PT, Borkhardt A, Surowy H, Wieczorek D, Meyer R, Eggermann T, Begemann M, Van Schaftingen E, Häusler M, Tenbrock K, van den Heuvel L, Elbracht M, Kurth, Kraft F. A human multisystem disorder

with autoinflammation, leukoencephalopathy and hepatopathy is caused by mutations in C2orf69. *J Clin Invest.* 131(12):e143078*,* 2021

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**Publications 2020**

345. Kölbel H, Roos A, van der Ven PFM, Evangelista T, Nolte K, Johnson K, Töpf A, Wilson M, Kress W, Sickmann A, Straub V, Kollipara L, **Weis J**, Fürst DO, Schara U. First clinical and myopathological description of a myofibrillar myopathy with congenital onset and homozygous mutation in FLNC. *Hum Mutat.* 41(9): 1600-1614, 2020

344. Rabenstein M, **Weis J**, Abicht A, Fink GR, Lehmann HC, Wunderlich G. [Multiple acyl-CoA dehydrogenase deficiency/glutaric aciduria type 2: difficult diagnosis, easy to treat]. *Nervenarzt.* 91(4): 349-352, 2020

343. Mair D, Biskup S, Kress W, Abicht A, Brück W, Zechel S, Knop KC, Koenig FB, Tey S, Nikolin S, Eggermann K, Kurth I, Ferbert A, **Weis J**. Differential diagnosis of vacuolar myopathies in the NGS era. *Brain Pathol.* 30(5): 877-896, 2020

342. Mathis S, Vallat JM, **Weis J**. When botany inspired pathology of the peripheral nervous system. *Neurology.* 95(12): 532-536, 2020

341. Hedberg-Oldfors C, Meyer R, Nolte K, Abdul Rahim Y, Lindberg C, Karason K, Thuestad IJ, Visuttijai K, Geijer M, Begemann M, Kraft F, Lausberg E, Hitpass L, Götzl R, Luna EJ, Lochmüller H, Koschmieder S, Gramlich M, Gess B, Elbracht M, **Weis J**, Kurth I, Oldfors A, Knopp C. Loss of supervillin causes myopathy with myofibrillar disorganization and autophagic vacuoles. *Brain.* 143(8): 2406-2420, 2020

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339. Yilmazer-Hanke D, Mayer T, Müller HP, Neugebauer H, Abaei A, Scheuerle A, **Weis J**, Forsberg KME, Althaus K, Meier J, Ludolph AC, Del Tredici K, Braak H, Kassubek J, Rasche V. Histological correlates of postmortem ultra-high-resolution single-section MRI in cortical cerebral microinfarcts. *Acta Neuropathol Commun.* 8(1): 33, 2020

338. Altinova H, Hammes S, Palm M, Achenbach P, Gerardo-Nava J, Deumens R, Führmann T, van Neerven SGA, Hermans E, **Weis J**, Brook G. Dense fibroadhesive scarring and poor blood vessel-maturation hamper the integration of implanted collagen scaffolds in an experimental model of spinal cord injury. *Biomed Mater.* 15(1): 015012, 2020

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333. Rudnik-Schöneborn S, Huemer M, **Weis J**, Sauer E, Meng G. Early onset facioscapulohumeral muscular dystrophy - Long-term follow-up of a patient with total facial diplegia. *Neuromuscul Disord.* 29(12): 973-976, 2019

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328. Ross JA, Levy Y, Ripolone M, Kolb JS, Turmaine M, Holt M, Lindqvist J, Claeys KG, **Weis J**, Monforte M, Tasca G, Moggio M, Figeac N, Zammit PS, Jungbluth H,Fiorillo C, Vissing J, Witting N, Granzier H, Zanoteli E, Hardeman EC, Wallgren-Pettersson C, Ochala J. Impairments in contractility and cytoskeletal organisation cause nuclear defects in nemaline myopathy. *Acta Neuropathol.* 138(3): 477-495, 2019

327. Fischbach F, Nedelcu J, Leopold P, Zhan J, Clarner T, Nellessen L, Beißel C, van Heuvel Y, Goswami A, **Weis J**, Denecke B, Schmitz C, Hochstrasser T, Nyamoya S, Victor M, Beyer C, Kipp M. Cuprizone-induced graded oligodendrocyte vulnerability is regulated by the transcription factor DNA damage-inducible transcript 3. *Glia.* 67(2): 263-276, 2019

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324. Karsai G, Kraft F, Haag N, Korenke GC, Hänisch B, Othman A, Suriyanarayanan S, Steiner R, Knopp C, Mull M, Bergmann M, Schröder JM, **Weis J**, Elbracht M, Begemann M, Hornemann T, Kurth I. DEGS1-associated aberrant sphingolipid metabolism impairs nervous system function in humans. *J Clin Invest.* 129(3): 1229-1239, 2019

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Tacke M, Gerstl L, Young P, Horvath R, Senderek J. SACS variants are a relevant cause of autosomal recessive hereditary motor and sensory neuropathy. *Hum Genet.* 137(11-12): 911-919, 2018

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