Curriculum Vitae

Personal information

Markus Weber

Work experience

Since 1/06-current Head Neuromuscular Diseases Center/ALS Clinic, Kantonsspital St.Gallen, Switzerland, Director of Unit

4/2012 Professorship in Neurology, University of Basel

2000-2006 Senior neurologist, Department of Neurology, Kantonsspital St.Gallen, Switzerland

6/97- 8/00 EMG/ Neuromuscular fellowship, Neuromuscular Diseases Unit, Vancouver Hospital, University of British Columbia, Canada,

5/96 - 6/97 Senior Neurologist Department of Neurology, University Hospital, Essen, Germany

Education and training

1988-1995 Residency in Gynecology, Neurology; Psychiatry (Siegen, Stuttgart, Göppingen), Germany; Board certified neurologist

1983 - 89 Medicine, Universities of Bonn and Tübingen, Germany; Graduation, Medical Doctor (M.D.)

1982 - 83 Psychology, University of Konstanz, Germany Skillls: Statistics

Additional information

Publications

to be updated

1. Shepheard SR, Parker MD, Cooper-Knock J, Verber NS, Tuddenham L, Heath P, Beauchamp N, Place E, Sollars ESA, Turner MR, Malaspina A, Fratta P, Hewamadduma C, Jenkins TM, McDermott CJ, Wang D, Kirby J, Shaw PJ, Project MC, Project Min E. Value of systematic genetic screening of patients with amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry 2021;92:510-518.

2. Moisse M, Zwamborn RAJ, van Vugt J, van der Spek R, van Rheenen W, Kenna B, Van Eijk K, Kenna K, Corcia P, Couratier P, Vourc'h P, Hardiman O, McLaughin R, Gotkine M, Drory V, Ticozzi N, Silani V, de Carvalho M, Mora Pardina JS, Povedano M, Andersen PM, **Weber** M, Basak NA, Chen X, Eberle MA, Al-Chalabi A, Shaw C, Shaw PJ, Morrison KE, Landers JE, Glass JD, Robberecht W, van Es M, van den Berg L, Veldink J, Van Damme P, Project Min ESC. The Effect of SMN Gene Dosage on ALS Risk and Disease Severity. Ann Neurol 2021;89:686-697.

3. van Eijk RPA, Kliest T, McDermott CJ, Roes KCB, Van Damme P, Chio A, **Weber** M, Ingre C, Corcia P, Povedano M, Reviers E, van Es MA, Al-Chalabi A, Hardiman O, van den Berg LH. TRICALS: creating a highway toward a cure. Amyotroph Lateral Scler Frontotemporal Degener 2020;21:496-501.

4. Nihei Y, Mori K, Werner G, Arzberger T, Zhou Q, Khosravi B, Japtok J, Hermann A, Sommacal A, **Weber** M, German Consortium for Frontotemporal Lobar D, Bavarian Brain Banking A, Kamp F, Nuscher B, Edbauer D, Haass C. Poly-glycine-alanine exacerbates C9orf72 repeat expansion-mediated DNA damage via sequestration of phosphorylated ATM and loss of nuclear hnRNPA3. Acta Neuropathol 2020;139:99-118.

5. Neuwirth C, **Weber** M. Estimating the Number of Motor Neurons: First Go to the Roots Before Cropping the Fruits. Muscle Nerve 2020;62:154-155.

6. Neuwirth C, **Weber** M. MUNIX in children with spinal muscular atrophy: An unexpected journey. Muscle Nerve 2020.

7. Haider A, Gobbi L, Kretz J, Ullmer C, Brink A, Honer M, Woltering TJ, Muri D, Iding H, Burkler M, Binder M, Bartelmus C, Knuesel I, Pacher P, Herde AM, Spinelli F, Ahmed H, Atz K, Keller C, **Weber** M, Schibli R, Mu L, Grether U, Ametamey SM. Identification and Preclinical Development of a 2,5,6-Trisubstituted Fluorinated Pyridine Derivative as a Radioligand for the Positron Emission Tomography Imaging of Cannabinoid Type 2 Receptors. J Med Chem 2020;63:10287-10306.

8. Braun N, Macklin EA, Sinani E, Sherman A, **Weber** M, Pooled Resource Open-Access ALSCTC. The revised El Escorial criteria "clinically probable laboratory supported ALS"-once a promising now a superfluous category? Amyotroph Lateral Scler Frontotemporal Degener 2020;21:24-28.

9. van den Berg LH, Sorenson E, Gronseth G, Macklin EA, Andrews J, Baloh RH, Benatar M, Berry JD, Chio A, Corcia P, Genge A, Gubitz AK, Lomen-Hoerth C, McDermott CJ, Pioro EP, Rosenfeld J, Silani V, Turner MR, **Weber** M, Brooks BR, Miller RG, Mitsumoto H, Airlie House ALSCTGG. Revised Airlie House consensus guidelines for design and implementation of ALS clinical trials. Neurology 2019;92:e1610-e1623.

10. Stampfli P, Sommer S, Czell D, Kozerke S, Neuwirth C, **Weber** M, Sartoretti-Schefer S, Seifritz E, Gutzeit A, Reischauer C. Investigation of Neurodegenerative Processes in Amyotrophic Lateral Sclerosis Using White Matter Fiber Density. Clin Neuroradiol 2019;29:493-503.

11. Stalberg E, van Dijk H, Falck B, Kimura J, Neuwirth C, Pitt M, Podnar S, Rubin DI, Rutkove S, Sanders DB, Sonoo M, Tankisi H, Zwarts M. Standards for quantification of EMG and neurography. Clin Neurophysiol 2019;130:1688-1729.

12. Muller Herde A, Schibli R, **Weber** M, Ametamey SM. Metabotropic glutamate receptor subtype 5 is altered in LPS-induced murine neuroinflammation model and in the brains of AD and ALS patients. Eur J Nucl Med Mol Imaging 2019;46:407-420.

13. Lingor P, **Weber** M, Camu W, Friede T, Hilgers R, Leha A, Neuwirth C, Gunther R, Benatar M, Kuzma-Kozakiewicz M, Bidner H, Blankenstein C, Frontini R, Ludolph A, Koch JC, Investigators R-A. ROCK-ALS: Protocol for a Randomized, Placebo-Controlled, Double-Blind Phase IIa Trial of Safety, Tolerability and Efficacy of the Rho Kinase (ROCK) Inhibitor Fasudil in Amyotrophic Lateral Sclerosis. Front Neurol 2019;10:293.

14. Johnsen B, Pugdahl K, Fuglsang-Frederiksen A, Kollewe K, Paracka L, Dengler R, Camdessanche JP, Nix W, Liguori R, Schofield I, Maderna L, Czell D, Neuwirth C, **Weber** M, Drory VE, Abraham A, Swash M, de Carvalho M. Diagnostic criteria for amyotrophic lateral sclerosis: A multicentre study of inter-rater variation and sensitivity. Clin Neurophysiol 2019;130:307-314.

15. Haider A, Kretz J, Gobbi L, Ahmed H, Atz K, Burkler M, Bartelmus C, Fingerle J, Guba W, Ullmer C, Honer M, Knuesel I, **Weber** M, Brink A, Herde AM, Keller C, Schibli R, Mu L, Grether U, Ametamey SM. Structure-Activity Relationship Studies of Pyridine-Based Ligands and Identification of a Fluorinated Derivative for Positron Emission Tomography Imaging of Cannabinoid Type 2 Receptors. J Med Chem 2019;62:11165-11181.

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17. Forsberg K, Graffmo K, Pakkenberg B, **Weber** M, Nielsen M, Marklund S, Brannstrom T, Andersen PM. Misfolded SOD1 inclusions in patients with mutations in C9orf72 and other ALS/FTD-associated genes. J Neurol Neurosurg Psychiatry 2019;90:861-869.

18. Czell D, Neuwirth C, **Weber** M, Sartoretti-Schefer S, Gutzeit A, Reischauer C. Nine Hole Peg Test and Transcranial Magnetic Stimulation: Useful to Evaluate Dexterity of the Hand and Disease Progression in Amyotrophic Lateral Sclerosis. Neurol Res Int 2019;2019:7397491.

19. Brenner D, Rosenbohm A, Yilmaz R, Muller K, Grehl T, Petri S, Meyer T, Grosskreutz J, Weydt P, Ruf W, Neuwirth C, **Weber** M, Pinto S, Claeys KG, Schrank B, Jordan B, Knehr A, Gunther K, Hubers A, Zeller D, Kubisch C, Jablonka S, Sendtner M, Klopstock T, de Carvalho M, Sperfeld A, Borck G, Volk AE, Dorst J, Weis J, Otto M, Schuster J, Del Tredici K, Braak H, Danzer KM, Freischmidt A, Meitinger T, Ludolph AC, Andersen PM, Weishaupt JH, German ALSnMNDNET. Reply: Adult-onset distal spinal muscular atrophy: a new phenotype associated with KIF5A mutations. Brain 2019;142:e67.

20. Alix JJP, Neuwirth C, Gelder L, Burkhardt C, Castro J, de Carvalho M, Gawel M, Goedee S, Grosskreutz J, Lenglet T, Moglia C, Omer T, Schrooten M, Nandedkar S, Stalberg E, Barkhaus PE, Furtula J, van Dijk JP, Baldinger R, Costa J, Otto M, Sandberg A, **Weber** M. Assessment of the reliability of the motor unit size index (MUSIX) in single subject "round-robin" and multi-centre settings. Clin Neurophysiol 2019;130:666-674.

21. Westeneng HJ, Debray TPA, Visser AE, van Eijk RPA, Rooney JPK, Calvo A, Martin S, McDermott CJ, Thompson AG, Pinto S, Kobeleva X, Rosenbohm A, Stubendorff B, Sommer H, Middelkoop BM, Dekker AM, van Vugt J, van Rheenen W, Vajda A, Heverin M, Kazoka M, Hollinger H, Gromicho M, Korner S, Ringer TM, Rodiger A, Gunkel A, Shaw CE, Bredenoord AL, van Es MA, Corcia P, Couratier P, **Weber** M, Grosskreutz J, Ludolph AC, Petri S, de Carvalho M, Van Damme P, Talbot K, Turner MR, Shaw PJ, Al-Chalabi A, Chio A, Hardiman O, Moons KGM, Veldink JH, van den Berg LH. Prognosis for patients with amyotrophic lateral sclerosis: development and validation of a personalised prediction model. Lancet Neurol 2018;17:423-433.

22. van der Spek RA, van Rheenen W, Pulit SL, Kenna KP, Ticozzi N, Kooyman M, McLaughlin RL, Moisse M, van Eijk KR, van Vugt J, Iacoangeli A, Andersen P, Nazli Basak A, Blair I, de Carvalho M, Chio A, Corcia P, Couratier P, Drory VE, Glass JD, Hardiman O, Mora JS, Morrison KE, Mitne-Neto M, Robberecht W, Shaw PJ, Panades MP, van Damme P, Silani V, Gotkine M, **Weber** M, van Es MA, Landers JE, Al-Chalabi A, van den Berg LH, Veldink JH, Project Mine Als Sequencing C. Reconsidering the causality of TIA1 mutations in ALS. Amyotroph Lateral Scler Frontotemporal Degener 2018;19:1-3.

23. Reischauer C, Gutzeit A, Neuwirth C, Fuchs A, Sartoretti-Schefer S, **Weber** M, Czell D. In-vivo evaluation of neuronal and glial changes in amyotrophic lateral sclerosis with diffusion tensor spectroscopy. Neuroimage Clin 2018;20:993-1000.

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C, Newhard C, Zakrzewski M, Rosier E, Hamel N, Raheja D, Raaijman J, Ferguson T, **Weber** M. Implementing Motor Unit Number Index (MUNIX) in a large clinical trial: Real world experience from 27 centres. Clin Neurophysiol 2018;129:1756-1762.

27. Nandedkar SD, Barkhaus PE, Stalberg EV, Neuwirth C, **Weber** M. Motor unit number index: Guidelines for recording signals and their analysis. Muscle Nerve 2018;58:374-380.

28. Maier M, Welt T, Wirth F, Montrasio F, Preisig D, McAfoose J, Vieira FG, Kulic L, Spani C, Stehle T, Perrin S, **Weber** M, Hock C, Nitsch RM, Grimm J. A human-derived antibody targets misfolded SOD1 and ameliorates motor symptoms in mouse models of amyotrophic lateral sclerosis. Sci Transl Med 2018;10.

29. Haider A, Spinelli F, Herde AM, Mu B, Keller C, Margelisch M, **Weber** M, Schibli R, Mu L, Ametamey SM. Evaluation of 4-oxo-quinoline-based CB2 PET radioligands in R6/2 chorea huntington mouse model and human ALS spinal cord tissue. Eur J Med Chem 2018;145:746-759.

30. Brenner D, Yilmaz R, Muller K, Grehl T, Petri S, Meyer T, Grosskreutz J, Weydt P, Ruf W, Neuwirth C, **Weber** M, Pinto S, Claeys KG, Schrank B, Jordan B, Knehr A, Gunther K, Hubers A, Zeller D, Kubisch C, Jablonka S, Sendtner M, Klopstock T, de Carvalho M, Sperfeld A, Borck G, Volk AE, Dorst J, Weis J, Otto M, Schuster J, Del Tredici K, Braak H, Danzer KM, Freischmidt A, Meitinger T, Strom TM, Ludolph AC, Andersen PM, Weishaupt JH, German ALSnMNDNET. Hot-spot KIF5A mutations cause familial ALS. Brain 2018;141:688-697.

31. Nordin A, Akimoto C, Wuolikainen A, Alstermark H, Forsberg K, Baumann P, Pinto S, de Carvalho M, Hubers A, Nordin F, Ludolph AC, Weishaupt JH, Meyer T, Grehl T, Schweikert K, **Weber** M, Burkhardt C, Neuwirth C, Holmoy T, Morita M, Tysnes OB, Benatar M, Wuu J, Lange DJ, Bisgard C, Asgari N, Tarvainen I, Brannstrom T, Andersen PM. Sequence variations in C9orf72 downstream of the hexanucleotide repeat region and its effect on repeat-primed PCR interpretation: a large multinational screening study. Amyotroph Lateral Scler Frontotemporal Degener 2017;18:256-264.

32. Neuwirth C, Barkhaus PE, Burkhardt C, Castro J, Czell D, de Carvalho M, Nandedkar S, Stalberg E, **Weber** M. Motor Unit Number Index (MUNIX) detects motor neuron loss in pre-symptomatic muscles in Amyotrophic Lateral Sclerosis. Clin Neurophysiol 2017;128:495-500.

33. Feddermann-Demont N, Junge A, **Weber** KP, Weller M, Dvorak J, Tarnutzer AA. Prevalence of potential sports-associated risk factors in Swiss amyotrophic lateral sclerosis patients. Brain Behav 2017;7:e00630.

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35. Burkhardt C, Neuwirth C, **Weber** M. Longitudinal assessment of the Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS): lack of practice effect in ALS patients? Amyotroph Lateral Scler Frontotemporal Degener 2017;18:202-209.

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38. van Rheenen W, Shatunov A, Dekker AM, McLaughlin RL, Diekstra FP, Pulit SL, van der Spek RA, Vosa U, de Jong S, Robinson MR, Yang J, Fogh I, van Doormaal PT, Tazelaar GH, Koppers M, Blokhuis AM, Sproviero W, Jones AR, Kenna KP, van Eijk KR, Harschnitz O, Schellevis RD, Brands WJ, Medic J, Menelaou A, Vajda A, Ticozzi N, Lin K, Rogelj B, Vrabec K, Ravnik-Glavac M, Koritnik B, Zidar J, Leonardis L, Groselj LD, Millecamps S, Salachas F, Meininger V, de Carvalho M, Pinto S, Mora JS, Rojas-Garcia R, Polak M, Chandran S, Colville S, Swingler R, Morrison KE, Shaw PJ, Hardy J, Orrell RW, Pittman A, Sidle K, Fratta P, Malaspina A, Topp S, Petri S, Abdulla S, Drepper C, Sendtner M, Meyer T, Ophoff RA, Staats KA, Wiedau-Pazos M, Lomen-Hoerth C, Van Deerlin VM, Trojanowski JQ, Elman L, McCluskey L, Basak AN, Tunca C, Hamzeiy H, Parman Y, Meitinger T, Lichtner P, Radivojkov-Blagojevic M, Andres CR, Maurel C, Bensimon G, Landwehrmeyer B, Brice A, Payan CA, Saker-Delye S, Durr A, Wood NW, Tittmann L, Lieb W, Franke A, Rietschel M, Cichon S, Nothen MM, Amouyel P, Tzourio C, Dartigues JF, Uitterlinden AG, Rivadeneira F, Estrada K, Hofman A, Curtis C, Blauw HM, van der Kooi AJ, de Visser M, Goris A, **Weber** M, Shaw CE, Smith BN, Pansarasa O, Cereda C, Del Bo R, Comi GP, D'Alfonso S, Bertolin C, Soraru G, Mazzini L, Pensato V, Gellera C, Tiloca C, Ratti A, Calvo A, Moglia C, Brunetti M, Arcuti S, Capozzo R, Zecca C, Lunetta C, Penco S, Riva N, Padovani A, Filosto M, Muller B, Stuit RJ, Registry P, Group S, Registry S, Consortium FS, Consortium S, Group NS, Blair I, Zhang K, McCann EP, Fifita JA, Nicholson GA, Rowe DB, Pamphlett R, Kiernan MC, Grosskreutz J, Witte OW, Ringer T, Prell T, Stubendorff B, Kurth I, Hubner CA, Leigh PN, Casale F, Chio A, Beghi E, Pupillo E, Tortelli R, Logroscino G, Powell J, Ludolph AC, Weishaupt JH, Robberecht W, Van Damme P, Franke L, Pers TH, Brown RH, Glass JD, Landers JE, Hardiman O, Andersen PM, Corcia P, Vourc'h P, Silani V, Wray NR, Visscher PM, de Bakker PI, van Es MA, Pasterkamp RJ, Lewis CM, Breen G, Al-Chalabi A, van den Berg LH, Veldink JH. Genome-wide association analyses identify new risk variants and the genetic architecture of amyotrophic lateral sclerosis. Nat Genet 2016;48:1043-1048.

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2015;10:34.

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49. Joyce PI, McGoldrick P, Saccon RA, **Weber** W, Fratta P, West SJ, Zhu N, Carter S, Phatak V, Stewart M, Simon M, Kumar S, Heise I, Bros-Facer V, Dick J, Corrochano S, Stanford MJ, Luong TV, Nolan PM, Meyer T, Brandner S, Bennett DL, Ozdinler PH, Greensmith L, Fisher EM, Acevedo-Arozena A. A novel SOD1-ALS mutation separates central and peripheral effects of mutant SOD1 toxicity. Hum Mol Genet 2015;24:1883-1897.

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Projects

Major scientific achievements

Main research interests cover outcome measures, clinical neurophysiology, trial designs and cannabinoid research. Our work was crucial in developing a motor unit estimation technique called MUNIX as a biomarker for ALS trials. As part of a JPND project (SOPHIA) 12 centres across Europe were trained in this technique. This method is now also used in clinical trials as a biomarker in early phase I and II clinical trials. As part of this we have trained and certified more than 40 centres in Europe, North America, Middle East and Asia

In collaboration with the ETH Zürich the first cannabinoid typ 2 radioligand has been developed, which is now available for clinical use. A clinical study on ALS will start in 2021.

My team has participated in several EU-funded projects (JPND, eRare) and has received numerous grants from national and international funding bodies and various foundations.

Our center is the only Swiss center that is a study site in phase I, II and III international clinical trials and participates in international scientific investigators-initiated research projects.

Memberships

Executive committee European ALS Consortium (EALSC)

Co-chair EAN Scientific Panel management group ALS/FTD

Board member Swiss ALS Foundation

Board member Remedys Foundation

Member research committee, European Neuromuscular Center (ENMC)

Other Relevant Information