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Academic Education with Graduation

1974 – 1979 Study of Biology at the University of Basel

Scientific Graduation

1979 – 1982 PhD in Biology, Biocenter, University of Basel
2000 Habilitation at the University of Freiburg

Professional Career

1983 - 1985 Postdoc at Institute of Biology III, Albert-Ludwigs-University Freiburg
1985 - 1992 Group leader in the Dept. of G. Köhler, Max-Planck-Inst. for Immunobiology
1992 - 2011 Head of Clinical Research Unit for Rheumatology. Medical Center - University of Freiburg
2010 Professor (apl) Albert-Ludwigs-University Freiburg
2010 - Group leader, Medical Center - University of Freiburg, Dept. of Rheumatology and Clinical Immunology and Center for Chronic Immunodeficiency

Other activities

2013 – 2015 Center for Chronic Immunodeficiency, board member
2013 – now Comprehensive Research Center TRR130, IRTG board member

2020 - now Editor of the section "B cell biology" in Frontiers in Immunology

Reviewer for Blood, Clinical and Experimental Immunology, European Journal of Immunology, Frontiers in Immunology, Journal of Allergy and Clinical Immunology, Journal of Immunology, and others.

Memberships

European Society for Immunodeficiencies ESID
German Society for Immunology DGfI

Grants

DFG: TRR130, TP 06 " BAFF-receptor function in B cell survival and immune responses" 2013 - 2021
DFG: Ei235/9-1 "Gene-edited lymphoid progenitors for adoptive transfers as treatment of primary immunodeficiencies", WP04 (e-rare) 2018 - 2021.

Peer-reviewed publications: 103

About our research group: we are studying the role of BAFF receptor (BAFFR)-dependent pro survival signals in regulating the life span and activation of human B cells applying a variety of cellular, molecular and approaches including the in vitro development and gene transfer and genetic inactivation in primary human B cells.

List of Publications

- 1) Ntellas, P, Dardiotis, E, Sevdali, E, Siokas, V, Aloizou, AM, Tsinti, G, Germenis, AE, Hadjigeorgiou, GM, , H and Speletas, M (2020). "TNFRSF13C/BAFFR P21R and H159Y polymorphisms in multiple sclerosis." Mult Scler Relat Disord **37**: 101422.
- 2) Wilhelm, I, Levit-Zerdoun, E, Jakob, J, Villringer, S, Frensch, M, Ubelhart, R, Landi, A, Muller, P, Imberty, A, Thuenauer, R, Claudinon, J, Jumaa, H, Reth, M, , H, Hobeika, E and Romer, W (2019). "Carbohydrate-dependent B cell activation by fucose-binding bacterial lectins." Sci Signal **12**(571).
- 3) Sevdali, E, Katsantoni, E, Smulski, CR, Moschovi, M, Palassopoulou, M, Kolokotsa, EN, Argentou, N, Giannakoulas, N, Adamaki, M, Vassilopoulos, G, Polychronopoulou, S, Germenis, AE, , H and Speletas, M (2019). "BAFF/APRIL System Is Functional in B-Cell Acute Lymphoblastic Leukemia in a Disease Subtype Manner." Front Oncol **9**: 594.
- 4) Smulski, CR and , H (2018). "BAFF and BAFF-Receptor in B Cell Selection and Survival." Front Immunol **9**: 2285.
- 5) Schubert, D, Klein, MC, Hassdenteufel, S, Caballero-Oteyza, A, Yang, L, Proietti, M, Bulashevskaya, A, Kemming, J, Kuhn, J, Winzer, S, Rusch, S, Fliegau, M, Schaffer, AA, Pfeffer, S, Geiger, R, Cavaliere, A, Cao, H, Yang, F, Li, Y, Rizzi, M, , H, Kobbe, R, Marks, AL, Peppers, BP, Hostoffer, RW, Puck, JM, Zimmermann, R and Grimbacher, B (2018). "Plasma cell deficiency in human subjects with heterozygous mutations in Sec61 translocon alpha 1 subunit (SEC61A1)." J Allergy Clin Immunol **141**(4): 1427-38.
- 6) Miggelbrink, AM, Logan, BR, Buckley, RH, Parrott, RE, Dvorak, CC, Kapoor, N, Abdel-Azim, H, Prockop, SE, Shyr, D, Decaluwe, H, Hanson, IC, Gillio, A, Davila Saldana, BJ, , H, Hopkins, G, Walter, JE, Whangbo, JS, Kohn, DB, Puck, JM, Cowan, MJ, Griffith, LM, Haddad, E, O'Reilly, RJ, Notarangelo, LD and Pai, SY (2018). "B-cell differentiation and IL-21 response in IL2RG/JAK3 SCID patients after hematopoietic stem cell transplantation." Blood **131**(26): 2967-77.
- 7) , H, Winkler, T and Ceredig, R (2018). "Editorial: Making Science Fun - A Tribute to Our Colleague and Friend, Prof. Antonius G. Rolink (1953-2017)." Front Immunol **9**: 2915.
- 8) van de Vosse, E, van Ostaijen-Ten Dam, MM, Vermaire, R, Verhard, EM, Waaijer, JL, Bakker, JA, Bernards, ST, , H, van Tol, MJ, van Dissel, JT and Haverkamp, MH (2017). "Recurrent respiratory tract infections (RRTI) in the elderly: A late onset mild immunodeficiency?" Clin Immunol **180**: 111-9.
- 9) Smulski, CR, Kury, P, Seidel, LM, Staiger, HS, Edinger, AK, Willen, L, Seidl, M, Hess, H, Salzer, U, Rolink, AG, Rizzi, M, Schneider, P and , H (2017). "BAFF- and TACI-Dependent Processing of BAFFR by ADAM Proteases Regulates the Survival of B Cells." Cell Rep **18**(9): 2189-202.
- 10) Smulski, CR, Decossas, M, Chekkat, N, Beyrath, J, Willen, L, Guichard, G, Lorenzetti, R, Rizzi, M, , H, Schneider, P and Fournel, S (2017). "Hetero-oligomerization between the TNF receptor superfamily members CD40, Fas and TRAILR2 modulate CD40 signalling." Cell Death Dis **8**(2): e2601.
- 11) Sic, H, Speletas, M, Cornacchione, V, Seidl, M, Be, M, Linghu, B, Yang, F, Sevdali, E, Germenis, AE, Oakeley, EJ, Vangrevelinghe, E, Sailer, AW, Traggiai, E, Gram, H and , H (2017). "An Activating Janus Kinase-3 Mutation Is Associated with Cytotoxic T Lymphocyte Antigen-4-Dependent Immune Dysregulation Syndrome." Front Immunol **8**: 1824.
- 12) Keller, B, Stumpf, I, Strohmeier, V, Usadel, S, Verhoeyen, E, , H and Warnatz, K (2017). "High SYK Expression Drives Constitutive Activation of CD21^{low} B Cells." J Immunol **198**(11): 4285-92.
- 13) Keller, B, Cseresnyes, Z, Stumpf, I, Wehr, C, Fliegau, M, Bulashevskaya, A, Usadel, S, Grimbacher, B, Rizzi, M, , H, Niesner, R and Warnatz, K (2017). "Disturbed canonical nuclear factor of kappa light chain signaling in B cells of patients with common variable immunodeficiency." J Allergy Clin Immunol **139**(1): 220-31 e8.
- 14) Voelxen, N, Wehr, C, Gutenberger, S, Keller, B, Erlacher, M, Dominguez-Conde, C, Bertele, D, Emerich, F, Pantic, M, Jennings, S, Rakhmanov, M, Foerster, C, Martens, UM, Platzbecker, U, Peter, HH, Fisch, P, Boztug, K, , H, Salzer, U and Warnatz, K (2016). "B-cell signaling in persistent polyclonal B lymphocytosis (PPBL)." Immunol Cell Biol **94**(9): 830-7.
- 15) Kienzler, AK and , H (2016). Human B cell development and tolerance. Encyclopedia of Immunobiology. M Ratcliffe, Elsevier Ltd. **1**: 105-21.
- 16) Janda, A, Schwarz, K, van der Burg, M, Vach, W, Ijspeert, H, Lorenz, MR, Elgizouli, M, Pieper, K, Fisch, P, Hagel, J, Lorenzetti, R, Seidl, M, Roesler, J, Hauck, F, Traggiai, E, Speckmann, C, Rensing-Ehl, A, Ehl, S, , H and Rizzi, M (2016). "Disturbed B-lymphocyte selection in autoimmune lymphoproliferative syndrome." Blood **127**(18): 2193-202.
- 17) Haerzschel, A, Catusse, J, Hutterer, E, Paunovic, M, Zirikli, K, , H, Krenn, PW, Hartmann, TN and

- Burger, M (2016). "BCR and chemokine responses upon anti-IgM and anti-IgD stimulation in chronic lymphocytic leukaemia." *Ann Hematol* **95**(12): 1979-88.
- 18) Xu, HC, Huang, J, Khairnar, V, Duhan, V, Pandyra, AA, Grusdat, M, Shinde, P, McIlwain, DR, Maney, SK, Gommerman, J, Lohning, M, Ohashi, PS, Mak, TW, Pieper, K, Sic, H, Speletas, M, , H, Ware, CF, Tumanov, AV, Kruglov, AA, Nedospasov, SA, Haussinger, D, Recher, M, Lang, KS and Lang, PA (2015). "Deficiency of the B cell-activating factor receptor results in limited CD169+ macrophage function during viral infection." *J Virol* **89**(9): 4748-59.
- 19) Stepensky, P, Rensing-Ehl, A, Gather, R, Revel-Vilk, S, Fischer, U, Nabhani, S, Beier, F, Brummen-dorf, TH, Fuchs, S, Zenke, S, Firat, E, Pessach, VM, Borkhardt, A, Rakhmanov, M, Keller, B, Warnatz, K, , H, Niedermann, G, Elpeleg, O and Ehl, S (2015). "Early-onset Evans syndrome, immunodeficiency, and premature immunosenescence associated with tripeptidyl-peptidase II deficiency." *Blood* **125**(5): 753-61.
- 20) , H (2015). Early B cell development. *Agammaglobulinemia*. A Plebani and V Lougaris, Springer International Publishing. **4**: 1 - 17.
- 21) Boisson, B, Laplantine, E, Dobbs, K, Cobat, A, Tarantino, N, Hazen, M, Lidov, HG, Hopkins, G, Du, L, Belkadi, A, Chrabieh, M, Itan, Y, Picard, C, Fournet, JC, , H, Tsitsikov, E, Pai, SY, Abel, L, Al-Herz, W, Casanova, JL, Israel, A and Notarangelo, LD (2015). "Human HOIP and LUBAC deficiency underlies autoinflammation, immunodeficiency, amylopectinosis, and lymphangiectasia." *J Exp Med* **212**(6): 939-51.
- 22) Willmann, KL, Klaver, S, Dogu, F, Santos-Valente, E, Gancarz, W, Bilic, I, Mace, E, Salzer, E, Conde, CD, Sic, H, Majek, P, Banerjee, PP, Vladimer, GI, Haskologlu, S, Bolkent, MG, Kupesiz, A, Condino-Neto, A, Colinge, J, Superti-Furga, G, Pickl, WF, van Zelm, MC, , H, Orange, JS, Ikin-ciogullari, A and Boztug, K (2014). "Biallelic loss-of-function mutation in NIK causes a primary immunodeficiency with multifaceted aberrant lymphoid immunity." *Nat Commun* **5**: 5360.
- 23) Sic, H, Kraus, H, Madl, J, Flittner, KA, von Munchow, AL, Pieper, K, Rizzi, M, Kienzler, AK, Ayata, K, Rauer, S, Kleuser, B, Salzer, U, Burger, M, Zirlik, K, Lougaris, V, Plebani, A, Romer, W, Loeffler, C, Scaramuzza, S, Villa, A, Noguchi, E, Grimbacher, B and , H (2014). "Sphingosine-1-phosphate receptors control B-cell migration through signaling components associated with primary immunodeficiencies, chronic lymphocytic leukemia, and multiple sclerosis." *J Allergy Clin Immunol* **134**(2): 420-8.
- 24) Sassi, A, Lazaroski, S, Wu, G, Haslam, SM, Fliegau, M, Mellouli, F, Patiroglu, T, Unal, E, Ozdemir, MA, Jouhadi, Z, Khadir, K, Ben-Khemis, L, Ben-Ali, M, Ben-Mustapha, I, Borchani, L, Pfeifer, D, Jakob, T, Khemiri, M, Asplund, AC, Gustafsson, MO, Lundin, KE, Falk-Sorqvist, E, Moens, LN, Gungor, HE, Engelhardt, KR, Dziadzio, M, Stauss, H, Fleckenstein, B, Meier, R, Prayitno, K, Maul-Pavicic, A, Schaffer, S, Rakhmanov, M, Henneke, P, Kraus, H, , H, Kolsch, U, Nadifi, S, Nilsson, M, Bejaoui, M, Schaffer, AA, Smith, CI, Dell, A, Barbouche, MR and Grimbacher, B (2014). "Hypomorphic homozygous mutations in phosphoglucomutase 3 (PGM3) impair immunity and increase serum IgE levels." *J Allergy Clin Immunol* **133**(5): 1410-9, 9 e1-13.
- 25) Salzer, E, Kansu, A, Sic, H, Majek, P, Ikin-ciogullari, A, Dogu, FE, Prengemann, NK, Santos-Valente, E, Pickl, WF, Bilic, I, Ban, SA, Kuloglu, Z, Demir, AM, Ensari, A, Colinge, J, Rizzi, M, , H and Boztug, K (2014). "Early-onset inflammatory bowel disease and common variable immunodeficiency-like disease caused by IL-21 deficiency." *J Allergy Clin Immunol* **133**(6): 1651-9 e12.
- 26) Rakhmanov, M, Sic, H, Kienzler, AK, Fischer, B, Rizzi, M, Seidl, M, Melkaoui, K, Unger, S, Moehle, L, Schmit, NE, Deshmukh, SD, Ayata, CK, Schuh, W, Zhang, Z, Cosset, FL, Verhoeyen, E, Peter, HH, Voll, RE, Salzer, U, , H and Warnatz, K (2014). "High levels of SOX5 decrease proliferative capacity of human B cells, but permit plasmablast differentiation." *PLoS One* **9**(6): e100328.
- 27) Pieper, K, Rizzi, M, Speletas, M, Smulski, CR, Sic, H, Kraus, H, Salzer, U, Fiala, GJ, Schamel, WW, Lougaris, V, Plebani, A, Hammarstrom, L, Recher, M, Germenis, AE, Grimbacher, B, Warnatz, K, Rolink, AG, Schneider, P, Notarangelo, LD and , H (2014). "A common single nucleotide polymorphism impairs B-cell activating factor receptor's multimerization, contributing to common variable immunodeficiency." *J Allergy Clin Immunol* **133**(4): 1222-5.
- 28) Kraus, H, Kaiser, S, Aumann, K, Bonelt, P, Salzer, U, Vestweber, D, Erlacher, M, Kunze, M, Burger, M, Pieper, K, Sic, H, Rolink, A, , H and Rizzi, M (2014). "A feeder-free differentiation system identifies autonomously proliferating B cell precursors in human bone marrow." *J Immunol* **192**(3): 1044-54.
- 29) , H, Kraus, H, Sic, H, Kienzler, AK and Rizzi, M (2014). "B cell biology: an overview." *Curr Allergy Asthma Rep* **14**(5): 434.

- 30) Decker, S, Finter, J, Forde, AJ, Kissel, S, Schwaller, J, Mack, TS, Kuhn, A, Gray, N, Follo, M, Jumaa, H, Burger, M, Zirlik, K, Pfeifer, D, Miduturu, CV, , H, Veelken, H and Dierks, C (2014). "PIM kinases are essential for chronic lymphocytic leukemia cell survival (PIM2/3) and CXCR4-mediated microenvironmental interactions (PIM1)." Mol Cancer Ther **13**(5): 1231-45.
- 31) Stepensky, P, Keller, B, Buchta, M, Kienzler, AK, Elpeleg, O, Somech, R, Cohen, S, Shachar, I, Miosge, LA, Schlesier, M, Fuchs, I, Enders, A, , H, Grimbacher, B and Warnatz, K (2013). "Deficiency of caspase recruitment domain family, member 11 (CARD11), causes profound combined immunodeficiency in human subjects." J Allergy Clin Immunol **131**(2): 477-85 e1.
- 32) Schena, F, Volpi, S, Faliti, CE, Penco, F, Santi, S, Proietti, M, Schenk, U, Damonte, G, Salis, A, Bellotti, M, Fais, F, Tenca, C, Gattorno, M, , H, Rizzi, M, Warnatz, K, Idzko, M, Ayata, CK, Rakhmanov, M, Galli, T, Martini, A, Canossa, M, Grassi, F and Traggiai, E (2013). "Dependence of immunoglobulin class switch recombination in B cells on vesicular release of ATP and CD73 ectonucleotidase activity." Cell Rep **3**(6): 1824-31.
- 33) Recher, M, Fried, AJ, Massaad, MJ, Kim, HY, Rizzini, M, Frugoni, F, Walter, JE, Mathew, D, , H, Hess, C, Giliani, S, Umetsu, DT, Notarangelo, LD and Geha, RS (2013). "Intronic SH2D1A mutation with impaired SAP expression and agammaglobulinemia." Clin Immunol **146**(2): 84-9.
- 34) Popadic, D, Anegon, I, Baeten, D, , H, Giese, T, Marits, P, Martinez-Caceres, E, Mascart, F, Nestle, F, Pujol-Borrell, R, Savic, E, Scheibenbogen, C, Seliger, B, Thunberg, S, Turina, M, Villanova, F, Winqvist, O and Wikstrom, AC (2013). "Predictive immunomonitoring -- the COST ENTIRE initiative." Clin Immunol **147**(1): 23-6.
- 35) Pieper, K, Grimbacher, B and , H (2013). "B-cell biology and development." J Allergy Clin Immunol **131**(4): 959-71.
- 36) Kienzler, AK, Rizzi, M, Reith, M, Nutt, SL and , H (2013). "Inhibition of human B-cell development into plasmablasts by histone deacetylase inhibitor valproic acid." J Allergy Clin Immunol **131**(6): 1695-9.
- 37) Cattaneo, F, Recher, M, Masneri, S, Baxi, SN, Fiorini, C, Antonelli, F, Wyszocki, CA, Calderon, JG, , H, Smith, AR, Bonilla, FA, Tsitsikov, E, Giliani, S, Notarangelo, LD and Pai, SY (2013). "Hypomorphic Janus kinase 3 mutations result in a spectrum of immune defects, including partial maternal T-cell engraftment." J Allergy Clin Immunol **131**(4): 1136-45.
- 38) Thiel, J, Kimmig, L, Salzer, U, Grudzien, M, Lebrecht, D, Hagena, T, Draeger, R, Voelxen, N, Bergbreiter, A, Jennings, S, Gutenberger, S, Aichem, A, Illges, H, Hannan, JP, Kienzler, AK, Rizzi, M, , H, Peter, HH, Warnatz, K, Grimbacher, B, Rump, JA and Schlesier, M (2012). "Genetic CD21 deficiency is associated with hypogammaglobulinemia." J Allergy Clin Immunol **129**(3): 801-10 e6.
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- 41) Rizzi, M, Neumann, C, Fielding, AK, Marks, R, Goldacker, S, Thaventhiran, J, Tarzi, MD, Schlesier, M, Salzer, U, , H, Warnatz, K, Finke, J, Grimbacher, B and Peter, HH (2011). "Outcome of allogeneic stem cell transplantation in adults with common variable immunodeficiency." J Allergy Clin Immunol **128**(6): 1371-4 e2.
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- 43) Ochtrop, ML, Goldacker, S, May, AM, Rizzi, M, Draeger, R, Hauschke, D, Stehfest, C, Warnatz, K, Goebel, H, Technau-Ihling, K, Werner, M, Salzer, U, , H, Schlesier, M and Peter, HH (2011). "T and B lymphocyte abnormalities in bone marrow biopsies of common variable immunodeficiency." Blood **118**(2): 309-18.
- 44) Janda, A, Rensing-Ehl, A, , H, Ehl, S and Speckmann, C (2011). "Elevated serum BAFF levels in

- patients with autoimmunity and lymphoproliferation." Scand J Immunol **74**(5): 518-9.
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 - 46) , H, Salzer, U and Warnatz, K (2010). "Common variable immunodeficiency at the end of a prospering decade: towards novel gene defects and beyond." Curr Opin Allergy Clin Immunol **10**(6): 526-33.
 - 47) Warnatz, K, Salzer, U, Rizzi, M, Fischer, B, Gutenberger, S, Bohm, J, Kienzler, AK, Pan-Hammarstrom, Q, Hammarstrom, L, Rakhmanov, M, Schlesier, M, Grimbacher, B, Peter, HH and , H (2009). "B-cell activating factor receptor deficiency is associated with an adult-onset antibody deficiency syndrome in humans." Proc Natl Acad Sci U S A **106**(33): 13945-50.
 - 48) Venhoff, N, Rizzi, M, Salzer, U, Bossaller, L, Thoden, J, , H and Walker, UA (2009). "Monozygotic twins with stiff person syndrome and autoimmune thyroiditis: rituximab inefficacy in a double-blind, randomised, placebo controlled crossover study." Ann Rheum Dis **68**(9): 1506-8.
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