Curriculum Vitae Prof. Dr. Marcus A. Mall

Professor und Direktor der Klinik für Pädiatrie m. S. Pneumologie, Immunologie und Intensivmedizin
Ärztlicher Centrumsleiter des CharitéCentrum 17 für Frauen-, Kinder- und Jugendmedizin mit Perinatalzentrum und Humangenetik
Charité - Universitätsmedizin Berlin

Akademischer und beruflicher Werdegang

Seit 2020	Ärztlicher Centrumsleiter des CharitéCentrum 17 für Frauen-, Kinder- und Jugendmedizin mit Perinatalzentrum und Humangenetik
Seit 2018	W3-Professur für Pädiatrische Pneumologie und Immunologie und Direktor der Klinik für Pädiatrie m. S. Pneumologie, Immunologie und Intensivmedizin & Mukoviszidose-Zentrum, Charité - Universitätsmedizin Berlin
Seit 2018	BIH-Professur, Berlin Institute of Health (BIH)
2012 – 2018	Direktor des Zentrums für Translationale Lungenforschung (TLRC) und der Abteilung für Translationale Pneumologie, Universität Heidelberg
2009 – 2018	W3-Professur und Leitung der Sektion für Pädiatrische Pneumologie & Allergologie und Mukoviszidose-Zentrum, Zentrum für Kinderheilkunde und Jugendmedizin, Universität Heidelberg
2009 – 2012	W3-Heisenberg-Professur für Translationale Pädiatrische Pneumologie
2007 – 2009	Oberarzt und Leiter des Mukoviszidose-Zentrums, Zentrum für Kinderheilkunde und Jugendmedizin, Universität Heidelberg
2007	Habilitation am Zentrum für Kinderheilkunde und Jugendmedizin, Universität Heidelberg
2006 – 2018	Gruppenleiter der Molecular Medicine Partnership Unit (MMPU) der Universität Heidelberg und des European Molecular Biology Laboratory (EMBL)
2006 – 2018	Fakultätsmitglied der Hartmut Hoffmann-Berling International Graduate School of Molecular & Cellular Biology Heidelberg (HBIGS)
2005 – 2009	Gruppenleiter der EU-geförderten Nachwuchsgruppe „Mukoviszidose/ Chronische Atemwegserkrankung“ (Marie Curie Excellence Team)
2004 – 2006	Wissenschaftlicher Mitarbeiter und Assistenzarzt, Zentrum für Kinderheilkunde und Jugendmedizin, Universität Heidelberg
2003 – 2004	Assistant Professor, Cystic Fibrosis/Pulmonary Research and Treatment Center, School of Medicine, University of North Carolina at Chapel Hill, Chapel Hill, NC, USA
2000 – 2003	Postdoc am Cystic Fibrosis/Pulmonary Research and Treatment Center, School of Medicine, University of North Carolina at Chapel Hill, Chapel Hill, NC, USA

1997 – 2000	Wissenschaftlicher Mitarbeiter und Assistenzarzt, Universitätskinderklinik, Universität Freiburg
1997	Promotion an der Universität Freiburg
1990 – 1997	Studium an der Universität Freiburg und am University College London School of Medicine (UCL), Vereinigtes Königreich

Klinische Qualifikationen

2017	Zusatzweiterbildung Allergologie
2011	Zusatzweiterbildung Infektiologie
2008	Zusatzweiterbildung Kinderpneumologie
2006	Facharzt für Kinder- und Jugendmedizin

Funktionen in wissenschaftlichen Gesellschaften und Gremien

2018 - 2020	Vize-Präsident der European Cystic Fibrosis Society (ECFS)
2017 - 2018	Chair und Co-Chair der ECFS Basic Science Conference
Seit 2017	Mitglied des Programmkomitees der North American Cystic Fibrosis Conference (NACFC)
2016	Tagungspräsident der 19. Deutschen Mukoviszidose Tagung, Würzburg
Seit 2015	Mitglied des wissenschaftlichen Beirats des SickKids-Cystic Fibrosis Canada Program for Individualized CF Therapy (CFIT), Toronto, Kanada
2014 -2020	Mitglied des Vorstands der European Cystic Fibrosis Society (ECFS)
2011	Vize-Präsident der 34. ECFS-Konferenz, Hamburg
2009 – 2012	Mitglied des ECFS Conference Scientific Steering Committee
2006 – 2012	Gewähltes Mitglied (2006-2009) und Vorsitzender (2010-2012) der Forschungsgemeinschaft Mukoviszidose (FGM) im Mukoviszidose e.V.

Projektkoordination, Mitgliedschaft in Verbundprojekten

Seit 2021	Koordinator des Standorts Berlin im Deutschen Zentrum für Kinder- und Jugendgesundheit (DZKJ)
Seit 2021	Stellvertretender Sprecher des SFB 1449 „Dynamische Hydrogele auf biologischen Grenzflächen“
Seit 2019	Konsortialführung und Sprecher des Innovationsfonds-Projekts „conneCT CF: Coaching und Telemonitoring für Patienten mit Cystischer Fibrose“
2018 - 2022	Projektleiter des SFB-TR 84 „Angeborene Immunität der Lunge: Mechanismen des Pathogenangriffs und der Wirtsabwehr in der Pneumonie"
2013-2016	Projektleiter des EU-Verbundprojekts CFMATTERS: Cystic Fibrosis Microbiome-determined Antimicrobial Therapy Trial in Exacerbations: Results Stratified (7. Rahmenprogramm)

Seit 2012	Sprecher des Krankheitsfelds Mukoviszidose im Deutsches Zentrum für Lungenforschung (DZL)
2011 – 2018	Gründungsdirektor und Vorstandsmitglied des Deutsches Zentrums für Lungenforschung (DZL)
2009 – 2015	Projektleiter des BMBF-Verbundprojekts CARPuD: Netzwerk zellbasierte Verfahren für seltene Lungenerkrankungen

Ehrungen & Auszeichnungen

2020	Adalbert-Czerny-Preis der Deutschen Gesellschaft für Kinder- und Jugendmedizin (DGKJ)
2020	ERS Excellence Award for Research in Cystic Fibrosis
2018	Einstein-Professur, Einstein-Stiftung Berlin
2017	Fellow of ERS (FERS)
2009	Heisenberg-Professur für Translationale Pädiatrische Pulmologie, Deutsche Forschungsgemeinschaft (DFG)
2009	Forschungspreis der Deutschen Gesellschaft für Pneumologie (DGP)
2005	Marie Curie Excellence Grant (EU 6. Rahmenprogramm))
2005	Johannes Wenner Preis der Deutschen Gesellschaft für Pädiatrische Pneumologie (GPP)
2005	Professor David Shmerling Forschungspreis, Deutsche Gesellschaft für Pädiatrische Gastroenterologie und Ernährung (GPGE)
2000	Forschungsstipendium, Deutsche Forschungsgemeinschaft (DFG)
1998	Albrecht Fleckenstein Nachwuchspreis der Universität Freiburg
1997	Adolf Windorfer Preis für Mukoviszidoseforschung, Mukoviszidoes e.V.
1994 – 1997	Stipendium der Studienstiftung des deutschen Volkes
1993 – 1994	Stipendium des Deutschen Akademischen Austauschdienstes (DAAD)

Mitgliedschaft in Fachgesellschaften

Seit 2019	Fleischner Society
Seit 2010	European Respiratory Society (ERS)
Seit 2010	Deutsche Gesellschaft für Pneumologie (DGP)
Seit 2006	American Thoracic Society (ATS)
Seit 2004	Bundesverband Cystische Fibrose (Mukoviszidose e.V.)
Seit 1999	European Cystic Fibrosis Society (ECFS)
Seit 1998	Deutsche Physiologische Gesellschaft (DPG)
Seit 1998	Deutsche Gesellschaft für Kinder- und Jugendmedizin (DGKJ)
Seit 1998	Gesellschaft für Pädiatrische Pneumologie (GPP)

Publikationen

Originalarbeiten

Loske J, Rohmel J, Lukassen S, Stricker S, Magalhaes VG, Liebig J, Chua RL, Thurmann L, Messingschlager M, Seegebarth A, Timmermann B, Klages S, Ralser M, Sawitzki B, Sander LE, Corman VM, Conrad C, Laudi S, Binder M*****, Trump S*****, Eils R*****, **Mall MA***, Lehmann I*****. Pre-activated antiviral innate immunity in the upper airways controls early SARS-CoV-2 infection in children. *Nat Biotechnol* 2021. [Epub ahead of print]. (*equal contribution)

Loyal L, Braun J, Henze L, Kruse B, Dingeldey M, Reimer U, Kern F, Schwarz T, Mangold M, Unger C, Dorfler F, Kadler S, Rosowski J, Gurcan K, Uyar-Aydin Z, Frentsch M, Kurth F, Schnatbaum K, Eckey M, Hippenstiel S, Hocke A, Muller MA, Sawitzki B, Miltenyi S, Paul F, **Mall MA**, Wenschuh H, Voigt S, Drosten C, Lauster R, Lachman N, Sander LE, Corman VM, Rohmel J, Meyer-Arndt L, Thiel A, Giesecke-Thiel C. Cross-reactive CD4(+) T cells enhance SARS-CoV-2 immune responses upon infection and vaccination. *Science* 2021. [Epub ahead of print].

Goss CH, Fajac I, Jain R, Seibold W, Gupta A, Hsu MC, Sutharsan S, Davies JC, **Mall MA**. Efficacy and safety of inhaled ENaC inhibitor BI 1265162 in patients with cystic fibrosis: BALANCE-CF 1 - a randomised, Phase II study. *Eur Respir J* 2021. [Epub ahead of print].

Sharma A, Thongrom B, Bhatia S, von Lospichl B, Addante A, Graeber SY, Lauster D, **Mall MA**, Gradzielski M, Haag R. Polyglycerol-Based Mucus-Inspired Hydrogels. *Macromol Rapid Commun* 2021:e2100303. [Epub ahead of print].

Schroder A, Lunding LP, Zissler UM, Vock C, Webering S, Ehlers JC, Orinska Z, Chaker A, Schmidt-Weber CB, Lang NJ, Schiller HB, **Mall MA**, Fehrenbach H, Dinarello CA, Wegmann M. IL-37 regulates allergic inflammation by counterbalancing pro-inflammatory IL-1 and IL-33. *Allergy* 2021. [Epub ahead of print].

Theuring S, Thielecke M, van Loon W, Hommes F, Hulso C, von der Haar A, Korner J, Schmidt M, Bohringer F, **Mall MA**, Rosen A, von Kalle C, Kirchberger V, Kurth T, Seybold J, Mockenhaupt FP, Group BS. SARS-CoV-2 infection and transmission in school settings during the second COVID-19 wave: a cross-sectional study, Berlin, Germany, November 2020. *Euro Surveill* 2021;26.

Barry PJ*****, **Mall MA***, Alvarez A, Colombo C, de Winter-de Groot KM, Fajac I, McBennett KA, McKone EF, Ramsey BW, Sutharsan S, Taylor-Cousar JL, Tullis E, Ahluwalia N, Jun LS, Moskowitz SM, Prieto-Centurion V, Tian S, Waltz D, Xuan F, Zhang Y, Rowe SM, Polineni D, Group VXS. Triple Therapy for Cystic Fibrosis Phe508del-Gating and -Residual Function Genotypes. *N Engl J Med* 2021;385:815-825. (*equal contribution)

Stahl M, Steinke E, Graeber SY, Joachim C, Seitz C, Kauczor HU, Eichinger M, Hammerling S, Sommerburg O, Wielputz MO*****, **Mall MA***. Magnetic Resonance Imaging Detects Progression of Lung Disease and Impact of Newborn Screening in Preschool Children with Cystic Fibrosis. *Am J Respir Crit Care Med* 2021. [Epub ahead of print]. (*equal contribution)

Rohrich M, Leitz D, Glatting FM, Wefers AK, Weinheimer O, Flechsig P, Kahn N, **Mall MA**, Giesel FL, Kratochwil C, Huber PE, von Deimling A, Heussel CP, Kauczor HU, Kreuter M, Haberkorn UA. Fibroblast Activation Protein specific PET/CT imaging in fibrotic interstitial lung diseases and lung cancer: a translational exploratory study. *J Nucl Med* 2021. [Epub ahead of print].

Meisel C, Akbil B, Meyer T, Lankes E, Corman VM, Staudacher O, Unterwalder N, Kolsch U, Drosten C, **Mall MA**, Kallinich T, Schnabel D, Goffinet C, von Bernuth H. Mild COVID-19 despite autoantibodies to type I IFNs in Autoimmune-Polyendocrine-Syndrome Type 1 (APS-1). *J Clin Invest* 2021;131:e150867.

Hagner M, Albrecht M, Guerra M, Braubach P, Halle O, Zhou-Suckow Z, Butz S, Jonigk D, Hansen G, Schultz C, Dittrich AM*, **Mall MA***. IL-17A from innate and adaptive lymphocytes contributes to inflammation and damage in cystic fibrosis lung disease. *Eur Respir J* 2021;57. (*equal contribution)

Gertler M, Krause E, van Loon W, Krug N, Kausch F, Rohardt C, Rossig H, Michel J, Nitsche A, **Mall MA**, Nikolai O, Hommes F, Burock S, Lindner AK, Mockenhaupt FP, Pison U, Seybold J. Self-collected oral, nasal and saliva samples yield sensitivity comparable to professionally collected oro-nasopharyngeal swabs in SARS-CoV-2 diagnosis among symptomatic outpatients. *Int J Infect Dis* 2021;110:261-266.

Engelmann TA, Knudsen L, Leitz DHW, Duerr J, Beers MF, **Mall MA**, Ochs M. Linking Fibrotic Remodeling and Ultrastructural Alterations of Alveolar Epithelial Cells after Deletion of Nedd4-2. *Int J Mol Sci* 2021;22.

Zemanick ET, Taylor-Cousar JL, Davies J, Gibson RL, **Mall MA**, McKone EF, McNally P, Ramsey BW, Rayment JH, Rowe SM, Tullis E, Ahluwalia N, Chu C, Ho T, Moskowitz SM, Noel S, Tian S, Waltz D, Weinstock TG, Xuan F, Wainwright CE, McColley SA. A Phase 3 Open-Label Study of Elexacaftor/Tezacaftor/Ivacaftor in Children 6 through 11 Years of Age with Cystic Fibrosis and at Least One F508del Allele. *Am J Respir Crit Care Med* 2021;203:1522-1532.

Leitz DHW, Duerr J, Mulugeta S, Agircan AS, Zimmermann S, Kawabe H, Dalpke AH, Beers MF, **Mall MA**. Congenital Deletion of Nedd4-2 in Lung Epithelial Cells Causes Progressive Alveolitis and Pulmonary Fibrosis in Neonatal Mice. *International Journal of Molecular Sciences* 2021;22:6146.

Graeber SY, Boutin S, Wielputz MO, Joachim C, Frey DL, Wege S, Sommerburg O, Kauczor HU, Stahl M, Dalpke AH, **Mall MA**. Effects of Lumacaftor-Ivacaftor on Lung Clearance Index, Magnetic Resonance Imaging, and Airway Microbiome in Phe508del Homozygous Patients with Cystic Fibrosis. *Ann Am Thorac Soc* 2021;18:971-980.

Thielecke M, Theuring S, van Loon W, Hommes F, **Mall MA**, Rosen A, Bohringer F, von Kalle C, Kirchberger V, Kurth T, Seybold J, Mockenhaupt FP, group Bs. SARS-CoV-2 infections in kindergartens and associated households at the start of the second wave in Berlin, Germany - a cross sectional study. *Eur J Public Health* 2021. [Epub ahead of print].

Frey DL, Guerra M, **Mall MA**, Schultz C. Monitoring Neutrophil Elastase and Cathepsin G Activity in Human Sputum Samples. *J Vis Exp* 2021.

Thee S, Stahl M, Fischer R, Sutharsan S, Ballmann M, Muller A, Lorenz D, Urbanski-Rini D, Puschner F, Amelung VE, Fuchs C, **Mall MA**. A multi-centre, randomized, controlled trial on coaching and telemonitoring in patients with cystic fibrosis: conneCT CF. *BMC Pulm Med* 2021;21:131.

Maschmeyer P, Heinz GA, Skopnik CM, Lutter L, Mazzoni A, Heinrich F, von Stuckrad SL, Wirth LE, Tran CL, Riedel R, Lehmann K, Sakwa I, Cimaz R, Giudici F, **Mall MA**, Enghard P, Vastert B, Chang HD, Durek P, Annunziato F, van Wijk F, Radbruch A, Kallinich T, Mashreghi

MF. Antigen-driven PD-1(+) TOX(+) BHLHE40(+) and PD-1(+) TOX(+) EOMES(+) T lymphocytes regulate juvenile idiopathic arthritis in situ. *Eur J Immunol* 2021;51:915-929.

van Loon W, Hommes F, Theuring S, von der Haar A, Korner J, Schmidt M, von Kalle C, **Mall MA**, Seybold J, Kurth T, Mockenhaupt FP. Renewed absence of SARS-CoV-2 infections in the day care context in Berlin, January 2021. *Clin Infect Dis* 2021. [Epub ahead of print].

Sommerburg O, Hammerling S, Schneider SP, Okun J, Langhans CD, Leutz-Schmidt P, Wielputz MO, Siems W, Graber SY, **Mall MA***, Stahl M*. CFTR Modulator Therapy with Lumacaftor/Ivacaftor Alters Plasma Concentrations of Lipid-Soluble Vitamins A and E in Patients with Cystic Fibrosis. *Antioxidants (Basel)* 2021;10. (*equal contribution)

Hommes F, van Loon W, Thielecke M, Abramovich I, Lieber S, Hammerich R, Gehrke-Beck S, Linzbach E, Schuster A, von dem Busche K, Theuring S, Gertler M, Martinez GE, Richter J, Bergmann C, Bolke A, Bohringer F, **Mall MA**, Rosen A, Krannich A, Keller J, Bethke N, Kurzmann M, Kurth T, Kirchberger V, Seybold J, Mockenhaupt FP, Study Group B. SARS-CoV-2 Infection, Risk Perception, Behaviour and Preventive Measures at Schools in Berlin, Germany, during the Early Post-Lockdown Phase: A Cross-Sectional Study. *Int J Environ Res Public Health* 2021;18.

Ferreira-Gomes M, Kruglov A, Durek P, Heinrich F, Tizian C, Heinz GA, Pascual-Reguant A, Du W, Mothes R, Fan C, Frischbutter S, Habenicht K, Budzinski L, Ninnemann J, Jani PK, Guerra GM, Lehmann K, Matz M, Ostendorf L, Heiberger L, Chang HD, Bauherr S, Maurer M, Schonrich G, Raftery M, Kallinich T, **Mall MA**, Angermair S, Treskatsch S, Dorner T, Corman VM, Diefenbach A, Volk HD, Elezkurtaj S, Winkler TH, Dong J, Hauser AE, Radbruch H, Witkowski M, Melchers F, Radbruch A, Mashreghi MF. SARS-CoV-2 in severe COVID-19 induces a TGF-beta-dominated chronic immune response that does not target itself. *Nat Commun* 2021;12:1961.

Brown R, Small DM, Doherty DF, Holsinger L, Booth R, Williams R, Ingram RJ, Elborn JS, **Mall MA**, Taggart CC, Weldon S. Therapeutic Inhibition of Cathepsin S Reduces Inflammation and Mucus Plugging in Adult betaENaC-Tg Mice. *Mediators Inflamm* 2021;2021:6682657.

Salomon JJ, Albrecht T, Graeber SY, Scheuermann H, Butz S, Schatterny J, Mairbaurl H, Baumann I, **Mall MA**. Chronic rhinosinusitis with nasal polyps is associated with impaired TMEM16A-mediated epithelial chloride secretion. *J Allergy Clin Immunol* 2021;147:2191-2201 e2192.

Roehmel JF, Ogese MO, Rohrbach A, **Mall MA***, Naisbitt DJ*. Drug allergy to CFTR modulator therapy associated with lumacaftor-specific CD4+ T lymphocytes. *J Allergy Clin Immunol* 2021;147:753-756. (*equal contribution)

Griese M, Costa S, Linnemann RW, **Mall MA**, McKone EF, Polineni D, Quon BS, Ringshausen FC, Taylor-Cousar JL, Withers NJ, Moskowitz SM, Daines CL. Safety and Efficacy of Elexacaftor/Tezacaftor/Ivacaftor for 24 Weeks or Longer in People with Cystic Fibrosis and One or More F508del Alleles: Interim Results of an Open-Label Phase 3 Clinical Trial. *Am J Respir Crit Care Med* 2021;203:381-385.

Frey DL, Boutin S, Dittrich SA, Graeber SY, Stahl M, Wege S, Herth FJF, Sommerburg O, Schultz C, **Mall MA***, Dalpke AH*. Relationship between airway dysbiosis, inflammation and lung function in adults with cystic fibrosis. *J Cyst Fibros* 2021. (*equal contribution) [Epub ahead of print].

Guerra M, Halls VS, Schatterny J, Hagner M, **Mall MA***, Schultz C*. Protease FRET Reporters Targeting Neutrophil Extracellular Traps. *J Am Chem Soc* 2020;60:813-822. (*equal contribution)

Graeber SY, van Mourik P, Vonk AM, Kruisselbrink E, Hirtz S, van der Ent CK, **Mall MA***, Beekman JM*. Comparison of organoid swelling and *in vivo* biomarkers of CFTR function to determine effects of lumacaftor-ivacaftor in patients with cystic fibrosis homozygous for the F508del mutation. *Am J Respir Crit Care Med* 2020;202:1589-1592. (*equal contribution)

Triphan SMF, Stahl M, Jobst BJ, Sommerburg O, Kauczor HU, Schenk JP, Alrajab A, Eichinger M, **Mall MA**, Wielpütz MO. Echo Time-Dependence of Observed Lung T(1) in Patients With Cystic Fibrosis and Correlation With Clinical Metrics. *J Magn Reson Imaging* 2020;52:1645-1654.

Braun J, Loyal L, Frentsch M, Wendisch D, Georg P, Kurth F, Hippenstiel S, Dingeldey M, Kruse B, Fauchere F, Baysal E, Mangold M, Henze L, Lauster R, **Mall MA**, Beyer K, Röhmel J, Voigt S, Schmitz J, Miltenyi S, Demuth I, Müller MA, Hocke A, Witzenrath M, Suttorp N, Kern F, Reimer U, Wenschuh H, Drosten C, Corman VM, Giesecke-Thiel C, Sander LE, Thiel A. SARS-CoV-2-reactive T cells in healthy donors and patients with COVID-19. *Nature* 2020;587:270-274.

Stahl M, Joachim C, Kirsch I, Uselmann T, Yu Y, Alfeis N, Berger C, Minso R, Rudolf I, Stolpe C, Bovermann X, Liboschik L, Steinmetz A, Tennhardt D, Dorfler F, Rohmel J, Unorji-Frank K, Ruckes-Nilges C, von Stoutz B, Naehrlich L, Kopp MV, Dittrich AM, Sommerburg O, **Mall MA**. Multicentre feasibility of multiple-breath washout in preschool children with cystic fibrosis and other lung diseases. *ERJ Open Res* 2020;6:00408-2020

Vega G, Guequén A, Philp AR, Gianotti A, Arzola L, Villalón M, Zegarra-Moran O, Galietta LJ, **Mall MA**, Flores CA. Lack of Kcnn4 improves mucociliary clearance in muco-obstructive lung disease. *JCI Insight* 2020;5:e140076.

Kurth F, Roennefarth M, Thibeault C, Corman VM, Muller-Redetzky H, Mittermaier M, Ruwwe-Glosenkamp C, Heim KM, Krannich A, Zvorc S, Schmidt S, Kretzler L, Dang-Heine C, Rose M, Hummel M, Hocke A, Hubner RH, Opitz B, **Mall MA**, Rohmel J, Landmesser U, Pieske B, Knauss S, Endres M, Spranger J, Mockenhaupt FP, Tacke F, Treskatsch S, Angermair S, Siegmund B, Spies C, Weber-Carstens S, Eckardt KU, Schurmann D, Uhrig A, Stegemann MS, Zoller T, Drosten C, Suttorp N, Witzenrath M, Hippenstiel S, von Kalle C, Sander LE. Studying the pathophysiology of coronavirus disease 2019: a protocol for the Berlin prospective COVID-19 patient cohort (Pa-COVID-19). *Infection* 2020;48:619-626

Terliesner N, Lesniowski D, Krasnikova A, Korte M, Terliesner M, **Mall MA**, Dittrich K. Geographical accessibility of pediatric inpatient, nephrology, and urology services in Europe. *Front Pediatr* 2020;8:395.

Stuart WD, Guo M, Fink-Baldauf IM, Coleman AM, Clancy JP, **Mall MA**, Lim FY, Brewington JJ, Maeda Y. CRISPRi-mediated functional analysis of lung disease-associated loci at non-coding regions. *NAR Genom Bioinform* 2020;2:lqaa036.

Stoler I, Freytag J, Orak B, Unterwalder N, Henning S, Heim K, von Bernuth H, Krüger R, Winkler S, Eschenhagen P, Seipelt E, **Mall MA**, Foell D, Kessel C, Wittkowski H, Kallinich T. Gene-dose effect of MEFV gain-of-function mutations determines *ex vivo* neutrophil activation in familial mediterranean fever. *Front Immunol* 2020;11:716.

Sommerburg O, Wielputz MO, Trame JP, Wuennemann F, Optazaite E, Stahl M, Puderbach MU, Kopp-Schneider A, Fritzsching E, Kauczor HU, Baumann I, **Mall MA***, Eichinger M*. MRI detects chronic rhinosinusitis in infants and preschool children with cystic fibrosis. *Ann Am Thorac Soc* 2020;17:714-723. (*equal contribution)

Kumar V, Agrawal R, Pandey A, Kopf S, Hoeffgen M, Kaymak S, Bandapalli OR, Gorbunova V, Seluanov A, **Mall MA**, Herzig S, Nawroth PP. Compromised DNA repair is responsible for diabetes-associated fibrosis. *EMBO J* 2020;39:e103477.

Montgomery ST, Frey DL, **Mall MA**, Stick SM, Kicic A. Rhinovirus infection is associated with airway epithelial cell necrosis and inflammation via interleukin-1 in young children with cystic fibrosis. *Front Immunol* 2020;11:596.

Hagner M, Frey DL, Guerra M, Dittrich AS, Halls VS, Wege S, Herth FJF, Schultz C, **Mall MA**. New method for rapid and dynamic quantification of elastase activity on sputum neutrophils from patients with cystic fibrosis using flow cytometry. *Eur Respir J* 2020;55:1902355.

Geurts MH, de Poel E, Amatngalim GD, Oka R, Meijers FM, Kruisselbrink E, van Mourik P, Berkers G, de Winter-de Groot KM, Michel S, Muilwijk D, Aalbers BL, Mullenders J, Boj SF, Suen SWF, Brunsveld JE, Janssens HM, **Mall MA**, Graeber SY, van Boxtel R, van der Ent CK, Beekman JM, Clevers H. CRISPR-Based Adenine Editors Correct Nonsense Mutations in a Cystic Fibrosis Organoid Biobank. *Cell Stem Cell* 2020;26:503-510 e507.

Duerr J, Leitz DHW, Szczygiel M, Dvornikov D, Fraumann SG, Kreutz C, Zadora PK, Seyhan Agircan A, Konietzke P, Engelmann TA, Hegermann J, Mulugeta S, Kawabe H, Knudsen L, Ochs M, Rotin D, Muley T, Kreuter M, Herth FJF, Wielputz MO, Beers MF, Klingmuller U, **Mall MA**. Conditional deletion of Nedd4-2 in lung epithelial cells causes progressive pulmonary fibrosis in adult mice. *Nat Commun* 2020;11:2012.

Anagnostopoulou P, Latzin P, Jensen R, Stahl M, Harper A, Yammine S, Usemann J, Foong RE, Spycher B, Hall GL, Singer F, Stanojevic S, **Mall MA**, Ratjen F, Ramsey KA. Normative data for multiple breath washout outcomes in school-aged caucasian children. *Eur Respir J* 2020;55:1901302.

Konietzke P, Mueller J, Wuennemann F, Wagner WL, Schenk JP, Alrajab A, Kauczor HU, Stahl M, **Mall MA**, Wielputz MO, Sommerburg O. The value of chest magnetic resonance imaging compared to chest radiographs with and without additional lung ultrasound in children with complicated pneumonia. *PLoS One* 2020;15:e0230252.

Brown R, Paulsen M, Schmidt S, Schatterny J, Frank A, Hirtz S, Delaney R, Doherty D, Hagner M, Taggart C, Weldon S, **Mall MA**. Lack of IL-1 receptor signaling reduces spontaneous airway eosinophilia in juvenile mice with muco-obstructive lung disease. *Am J Respir Cell Mol Biol* 2020;62:300–309

Sörensen M, Kantorek J, Byrnes L, Boutin S, **Mall MA**, Lasitschka F, Zabeck H, Nguyen D, Dalpke AH. Pseudomonas aeruginosa modulates the antiviral response of bronchial epithelial cells. *Front Immunol* 2020;11:96.

Pieper M, Schulz-Hildebrandt H, **Mall MA***, Huttmann G*, Konig P*. Intravital microscopic optical coherence tomography imaging to assess mucus-mobilizing interventions for muco-obstructive lung disease in mice. Am J Physiol Lung *Cell Mol Physiol* 2020;318:L518-L524. (*equal contribution)

Heijerman HGM, McKone EF, Downey DG, Van Braeckel E, Rowe SM, Tullis E, **Mall MA**, Welter JJ, Ramsey BW, McKee CM, Marigowda G, Moskowitz SM, Waltz D, Sosnay PR, Simard C, Ahluwalia N, Xuan F, Zhang Y, Taylor-Cousar JL, McCoy KS. Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: A double-blind, randomised, phase 3 trial. *Lancet* 2019; 394: 1940-1948.

Middleton PG*, **Mall MA***, Dřevínek P, Lands LC, McKone EF, Polineni D, Ramsey BW, Taylor-Cousar JL, Tullis E, Vermeulen F, Marigowda G, McKee CM, Moskowitz SM, Nair N, Savage J, Simard C, Tian S, Waltz D, Xuan F, Rowe SM, Jain R. Elexacaftor–tezacaftor–ivacaftor for cystic fibrosis with a single Phe508del allele. *New England Journal of Medicine* 2019. (*equal contribution)

Belard S, Brand J, Schulze-Sturm U, Janda A, von Both U, Tacoli C, Alberer M, Kempf C, Stegemann MS, Kruger R, Varnholt V, Blohm M, Reiter K, Zoller T, Suttorp N, Mall M, von Bernuth H, Gratopp A, Hubner J, Hufnagel M, Kobbe R, Kurth F. Intravenous artesunate for imported severe malaria in children treated in four tertiary care centers in germany: A retrospective study. *Pediatr Infect Dis J* 2019; 38: e295-e300.

Sermet-Gaudelus I, Clancy JP, Nichols DP, Nick JA, De Boeck K, Solomon GM, **Mall MA**, Bolognese J, Bouisset F, den Hollander W, Paquette-Lamontagne N, Tomkinson N, Henig N, Elborn JS, Rowe SM. Antisense oligonucleotide eluforsen improves CFTR function in f508del cystic fibrosis. *J Cyst Fibros* 2019; 18: 536-542.

Wielputz MO, Eichinger M, Wege S, Eberhardt R, **Mall MA**, Kauczor HU, Puderbach MU, Risse F, Heussel CP, Heussel G. Midterm reproducibility of chest magnetic resonance imaging in adults with clinically stable cystic fibrosis and chronic obstructive pulmonary disease. *Am J Respir Crit Care Med* 2019; 200: 103-107.

Stahl M, Wielputz MO, Ricklefs I, Dopfer C, Barth S, Schlegtendal A, Graeber SY, Sommerburg O, Diekmann G, Husing J, Koerner-Rettberg C, Nahrlich L, Dittrich AM, Kopp MV, **Mall MA**. Preventive inhalation of hypertonic saline in infants with cystic fibrosis (PRESIS). A randomized, double-blind, controlled study. *Am J Respir Crit Care Med* 2019; 199: 1238-1248.

Stahl M, Joachim C, Wielputz MO, **Mall MA**. Comparison of lung clearance index determined by washout of N2 and SF6 in infants and preschool children with cystic fibrosis. *J Cyst Fibros* 2019; 18: 399-406.

Taylor-Cousar JL, **Mall MA**, Ramsey BW, McKone EF, Tullis E, Marigowda G, McKee CM, Waltz D, Moskowitz SM, Savage J, Xuan F, Rowe SM. Clinical development of triple-combination CFTR modulators for cystic fibrosis patients with one or two F508del alleles. *ERJ Open Res* 2019; 5.

Margaroli C, Garratt LW, Horati H, Dittrich AS, Rosenow T, Montgomery ST, Frey DL, Brown MR, Schultz C, Guglani L, Kicic A, Peng L, Scholte BJ, **Mall MA***, Janssens HM*, Stick SM*, Tirouvanziam R*. Elastase exocytosis by airway neutrophils is associated with early lung damage in children with cystic fibrosis. *Am J Respir Crit Care Med* 2019; 199: 873-881. (*equal contribution)

Small DM, Brown RR, Doherty DF, Abladey A, Zhou-Suckow Z, Delaney RJ, Kerrigan L, Dougan CM, Borensztajn KS, Holsinger L, Booth R, Scott CJ, Lopez-Campos G, Elborn JS,

Mall MA, Weldon S, Taggart CC. Targeting of cathepsin s reduces cystic fibrosis-like lung disease. *Eur Respir J* 2019; 53: 1801523.

Guerra M, Frey D, Hagner M, Dittrich S, Paulsen M, **Mall MA**, Schultz C. Cathepsin g activity as a new marker for detecting airway inflammation by microscopy and flow cytometry. *ACS Cent Sci* 2019; 5: 539-548.

Sommburg O, Helling-Bakki A, Alrajab A, Schenk JP, Springer W, **Mall MA**, Loukanov T, Eichhorn JG. Assessment of suspected vascular rings and slings and/or airway pathologies using magnetic resonance imaging rather than computed tomography. *Respiration* 2019; 97: 108-118.

Montgomery ST, Dittrich AS, Garratt LW, Turkovic L, Frey DL, Stick SM, **Mall MA***, Kicic A*. Interleukin-1 is associated with inflammation and structural lung disease in young children with cystic fibrosis. *J Cyst Fibros* 2018; 17: 715-722. (*equal contribution)

Keating D, Marigowda G, Burr L, Daines C, **Mall MA**, McKone EF, Ramsey BW, Rowe SM, Sass LA, Tullis E, McKee CM, Moskowitz SM, Robertson S, Savage J, Simard C, Van Goor F, Waltz D, Xuan F, Young T, Taylor-Cousar JL. Vx-445-tezacaftor-ivacaftor in patients with cystic fibrosis and one or two Phe508del alleles. *N Engl J Med* 2018; 379: 1612-1620.

Davies JC, Moskowitz SM, Brown C, Horsley A, **Mall MA**, McKone EF, Plant BJ, Prais D, Ramsey BW, Taylor-Cousar JL, Tullis E, Uluer A, McKee CM, Robertson S, Shilling RA, Simard C, Van Goor F, Waltz D, Xuan F, Young T, Rowe SM. Vx-659-tezacaftor-ivacaftor in patients with cystic fibrosis and one or two Phe508del alleles. *N Engl J Med* 2018; 379: 1599-1611.

Hahn A, Salomon JJ, Leitz D, Feigenbutz D, Korsch L, Lisewski I, Schrimpf K, Millar-Buchner P, **Mall MA**, Frings S, Mohrlen F. Expression and function of anoctamin 1/tmem16a calcium-activated chloride channels in airways of in vivo mouse models for cystic fibrosis research. *Pflugers Arch* 2018; 470: 1335-1348.

Wielputz MO, von Stackelberg O, Stahl M, Jobst BJ, Eichinger M, Puderbach MU, Nahrlich L, Barth S, Schneider C, Kopp MV, Ricklefs I, Buchholz M, Tummler B, Dopfer C, Vogel-Claussen J, Kauczor HU, **Mall MA**. Multicentre standardisation of chest mri as radiation-free outcome measure of lung disease in young children with cystic fibrosis. *J Cyst Fibros* 2018; 17: 518-527.

Graeber SY, Dopfer C, Naehrlich L, Gyulumyan L, Scheuermann H, Hirtz S, Wege S, Mairbaurl H, Dorda M, Hyde R, Bagheri-Hanson A, Rueckes-Nilges C, Fischer S, **Mall MA***, Tummler B*. Effects of lumacaftor-ivacaftor therapy on cystic fibrosis transmembrane conductance regulator function in Phe508del homozygous patients with cystic fibrosis. *Am J Respir Crit Care Med* 2018; 197: 1433-1442. (*equal contribution)

Boutin S, Weitnauer M, Hassel S, Graeber SY, Stahl M, Dittrich AS, **Mall MA**, Dalpke AH. One time quantitative pcr detection of pseudomonas aeruginosa to discriminate intermittent from chronic infection in cystic fibrosis. *J Cyst Fibros* 2018; 17: 348-355.

Balazs A, Balla Z, Kui B, Maleth J, Rakonczay Z, Jr., Duerr J, Zhou-Suckow Z, Schatterny J, Sendler M, Mayerle J, Kuhn JP, Tiszlavicz L, **Mall MA**, Hegyi P. Ductal mucus obstruction and reduced fluid secretion are early defects in chronic pancreatitis. *Front Physiol* 2018; 9: 632.

Leutz-Schmidt P, Stahl M, Sommerburg O, Eichinger M, Puderbach MU, Schenk JP, Alrajab A, Triphan SMF, Kauczor HU, **Mall MA**, Wielputz MO. Non-contrast enhanced magnetic

resonance imaging detects mosaic signal intensity in early cystic fibrosis lung disease. *Eur J Radiol* 2018; 101: 178-183.

Konietzke P, Weinheimer O, Wielputz MO, Savage D, Ziyeh T, Tu C, Newman B, Galban CJ, **Mall MA**, Kauczor HU, Robinson TE. Validation of automated lobe segmentation on paired inspiratory-expiratory chest CT in 8-14 year-old children with cystic fibrosis. *PLoS One* 2018; 13: e0194557.

Dittrich AS, Kuhbandner I, Gehrig S, Rickert-Zacharias V, Twigg M, Wege S, Taggart CC, Herth F, Schultz C, **Mall MA**. Elastase activity on sputum neutrophils correlates with severity of lung disease in cystic fibrosis. *Eur Respir J* 2018; 51.

Stahl M, Graeber SY, Joachim C, Barth S, Ricklefs I, Diekmann G, Kopp MV, Naehrlich L, **Mall MA**. Three-center feasibility of lung clearance index in infants and preschool children with cystic fibrosis and other lung diseases. *J Cyst Fibros* 2018; 17: 249-255.

Livraghi-Butrico A, Wilkinson KJ, Volmer AS, Gilmore RC, Rogers TD, Caldwell RA, Burns KA, Esther CR, Jr., **Mall MA**, Boucher RC, O'Neal WK, Grubb BR. Lung disease phenotypes caused by overexpression of combinations of alpha-, beta-, and gamma-subunits of the epithelial sodium channel in mouse airways. *Am J Physiol Lung Cell Mol Physiol* 2018; 314: L318-l331.

Boutin S, Depner M, Stahl M, Graeber SY, Dittrich AS, Legatzki A, von Mutius E*, **Mall M***, Dalpke A*. Comparison of oropharyngeal microbiota from children with asthma and cystic fibrosis. *Mediators Inflamm* 2017;2017:5047403. (*****equal contribution)

Stanke F, Hector A, Hedtfeld S, Hartl D, Griese M, Tummler B, **Mall MA**. An informative intragenic microsatellite marker suggests the IL-1 receptor as a genetic modifier in cystic fibrosis. *Eur Respir J* 2017;50:1700426.

Boutin S, Graeber SY, Stahl M, Dittrich AS, **Mall MA***, Dalpke AH*****. Chronic but not intermittent infection with Pseudomonas aeruginosa is associated with global changes of the lung microbiome in cystic fibrosis. *Eur Respir J* 2017;50:1701086. (*****equal contribution)

Kumar V, Fleming T, Terjung S, Gorzelanny C, Gebhardt C, Agrawal R, **Mall MA**, Ranzinger J, Zeier M, Madhusudhan T, Ranjan S, Isermann B, Liesz A, Deshpande D, Haring HU, Biswas SK, Reynolds PR, Hammes HP, Peperkok R, Angel P, Herzig S, Nawroth PP. Homeostatic nuclear RAGE-ATM interaction is essential for efficient DNA repair. *Nucleic Acids Res* 2017;45:10595-10613.

Hahn A, Faulhaber J, Srisawang L, Stortz A, Salomon JJ, **Mall MA**, Frings S, Mohrlen F. Cellular distribution and function of ion channels involved in transport processes in rat tracheal epithelium. *Physiol Rep* 2017;5:e13290.

Guo M, Tomoshige K, Meister M, Muley T, Fukazawa T, Tsuchiya T, Karns R, Warth A, Fink-Baldauf IM, Nagayasu T, Naomoto Y, Xu Y, **Mall MA**, Maeda Y. Gene signature driving invasive mucinous adenocarcinoma of the lung. *EMBO Mol Med* 2017;9:462-481.

Buschle LR, Kurz FT, Kampf T, Wagner WL, Duerr J, Stiller W, Konietzke P, Wunnemann F, **Mall MA**, Wielputz MO, Schlemmer HP, Ziener CH. Dephasing and diffusion on the alveolar surface. *Phys Rev E* 2017;95:022415.

Neves J, Leitz D, Kraut S, Brandenberger C, Agrawal R, Weissmann N, Muhlfeld C, **Mall MA***, Altamura S*, Muckenthaler MU*. Disruption of the Hepcidin/Ferroportin Regulatory

System Causes Pulmonary Iron Overload and Restrictive Lung Disease. *EBioMedicine* 2017;20:230-239. (*****equal contribution)

Sommerburg O, Stahl M, Hammermann J, Okun JG, Kulozik A, Hoffmann G, **Mall M**. [Newborn Screening on Cystic Fibrosis in Germany: Comparison of the new Screening Protocol with an Alternative Protocol]. *Klin Padiatr* 2017;229:59-66.

Stahl M, Wielputz MO, Graeber SY, Joachim C, Sommerburg O, Kauczor HU, Puderbach M, Eichinger M, **Mall MA**. Comparison of lung clearance index and magnetic resonance imaging for assessment of lung disease in children with cystic fibrosis. *Am J Respir Crit Care Med* 2017;195:349-359.

Fritzsching B, Hagner M, Dai L, Christochowitz S, Agrawal R, Van Bodegom C, Schmidt S, Schatterny J, Hirtz S, Brown R, Goritzka M, Duerr J, Zhou-Suckow Z, **Mall MA**. Impaired mucus clearance exacerbates allergen-induced type 2 airway inflammation in juvenile mice. *J Allergy Clin Immunol* 2017;140:190-203.

Weidler S, Stopsack KH, Hammermann J, Sommerburg O, **Mall MA**, Hoffmann GF, Kohlmuller D, Okun JG, Macek M, Jr., Votava F, Krulisova V, Balascakova M, Skalicka V, Lee-Kirsch MA, Stopsack M. A product of immunoreactive trypsinogen and pancreatitis-associated protein as second-tier strategy in cystic fibrosis newborn screening. *J Cyst Fibros* 2016;15:752-758.

Jia J, Conlon TM, Ballester LC, Seimetz M, Bednorz M, Zhou-Suckow Z, Weissmann N, Eickelberg O, **Mall MA***, Yildirim AO*. Cigarette smoke causes acute airway disease and exacerbates chronic obstructive lung disease in neonatal mice. *Am J Physiol Lung Cell Mol Physiol* 2016;311:L602-L610. (*****equal contribution)

Stahr CS, Samarage CR, Donnelley M, Farrow N, Morgan KS, Zosky G, Boucher RC, Siu KK, **Mall MA**, Parsons DW, Dubsky S, Fouras A. Quantification of heterogeneity in lung disease with image-based pulmonary function testing. *Sci Rep* 2016;6:29438.

Forde E, Schutte A, Reeves E, Greene C, Humphreys H, **Mall M**, Fitzgerald-Hughes D, Devocelle M. Differential In Vitro and In Vivo Toxicities of Antimicrobial Peptide Prodrugs for Potential Use in Cystic Fibrosis. *Antimicrob Agents Chemother* 2016;60:2813-2821.

Salomon JJ, Spahn S, Wang X, Fullekrug J, Bertrand CA, **Mall MA**. Generation and functional characterization of epithelial cells with stable expression of SLC26A9 Cl^- channels. *Am J Physiol Lung Cell Mol Physiol* 2016;310:L593-L602.

Oltmanns U, Palmowski K, Wielputz M, Kahn N, Baroke E, Eberhardt R, Wege S, Wiebel M, Kreuter M, Herth FJ, **Mall MA**. Optical coherence tomography detects structural abnormalitiesof the nasal mucosa in patients with cystic fibrosis. *J Cyst Fibros* 2016;15:216-222.

Kreuter M, Herth FJ, Wacker M, Leidl R, Hellmann A, Pfeifer M, Behr J, Witt S, Kauschka D, **Mall M**, Gunther A, Markart P. Exploring Clinical and Epidemiological Characteristics of Interstitial Lung Diseases: Rationale, Aims, and Design of a Nationwide Prospective Registry-The EXCITING-ILD Registry. *Biomed Res Int* 2015;2015:123876.

Graeber SY, Hug MJ, Sommerburg O, Hirtz S, Hentschel J, Heinzmann A, Dopfer C, Schulz A, Mainz JG, Tummler B*, **Mall MA***. Intestinal Current Measurements Detect Activation of Mutant CFTR in Patients with Cystic Fibrosis with the G551D Mutation Treated with Ivacaftor. *Am J Respir Crit Care Med* 2015;192:1252-1255. (*****equal contribution)

Oglesby IK, Vencken SF, Agrawal R, Gaughan K, Molloy K, Higgins G, McNally P, McElvaney NG, **Mall MA**, Greene CM. miR-17 overexpression in cystic fibrosis airway epithelial cells decreases interleukin-8 production. *Eur Respir J* 2015;46:1350-1360.

Galluzzo M, Ciraolo E, Lucattelli M, Hoxha E, Ulrich M, Campa CC, Lungarella G, Doring G, Zhou-Suckow Z, **Mall M**, Hirsch E, De Rose, V. Genetic Deletion and Pharmacological Inhibition of PI3K gamma Reduces Neutrophilic Airway Inflammation and Lung Damage in Mice with Cystic Fibrosis-Like Lung Disease. *Mediators Inflamm* 2015;2015:545417.

Marcos V, Zhou-Suckow Z, Onder YA, Bohla A, Hector A, Vitkov L, Krautgartner WD, Stoiber W, Griese M, Eickelberg O, **Mall MA**, Hartl D. Free DNA in cystic fibrosis airway fluids correlates with airflow obstruction. *Mediators Inflamm* 2015;2015:408935.

Seys LJ, Verhamme FM, Dupont LL, Desauter E, Duerr J, Seyhan AA, Conickx G, Joos GF, Brusselle GG, **Mall MA***, Bracke KR*. Airway Surface Dehydration Aggravates Cigarette Smoke-Induced Hallmarks of COPD in Mice. *PLoS ONE* 2015;10:e0129897. (*****equal contribution)

Vicuna L, Strochlic DE, Latremoliere A, Bali KK, Simonetti M, Husainie D, Prokosch S, Riva P, Griffin RS, Njoo C, Gehrig S, **Mall MA**, Arnold B, Devor M, Woolf CJ, Liberles SD, Costigan M, Kuner R. The serine protease inhibitor SerpinA3N attenuates neuropathic pain by inhibiting T cell-derived leukocyte elastase. *Nat Med* 2015;21:518-523.

Sommerburg O, De Spirt S, Mattern A, Joachim C, Langhans CD, Nesaretnam K, Siems W, Stahl W, **Mall MA**. Supplementation with red palm oil increases beta-carotene and vitamin A blood levels in patients with cystic fibrosis. *Mediators Inflamm* 2015;2015:817127.

Reunert J, Lotz-Havla AS, Polo G, Kannenberg F, Fobker M, Griese M, Mengel E, Muntau AC, Schnabel P, Sommerburg O, Borggraefe I, Dardis A, Burlina AP, **Mall MA**, Ciana G, Bembi B, Burlina AB, Marquardt T. Niemann-Pick Type C-2 Disease: Identification by Analysis of Plasma Cholestane-3beta,5alpha,6beta-Triol and Further Insight into the Clinical Phenotype. *JIMD Rep* 2015;23:17-26.

Sommerburg O, Hammermann J, Lindner M, Stahl M, Muckenthaler M, Kohlmueller D, Happich M, Kulozik AE, Stopsack M, Gahr M, Hoffmann GF*, **Mall MA***. Five years of experience with biochemical cystic fibrosis newborn screening based on IRT/PAP in Germany. *Pediatr Pulmonol* 2015;50:655-664. (*****equal contribution)

Fritzsching B, Zhou-Suckow Z, Trojanek JB, Schubert SC, Schatterny J, Hirtz S, Agrawal R, Muley T, Kahn N, Sticht C, Gunkel N, Welte T, Randell SH, Langer F, Schnabel P, Herth FJ, **Mall MA**. Hypoxic epithelial necrosis triggers neutrophilic inflammation via IL-1 receptor signaling in cystic fibrosis lung disease. *Am J Respir Crit Care Med* 2015;191:902-913.

Boutin S, Graeber SY, Weitnauer M, Panitz J, Stahl M, Clausznitzer D, Kaderali L, Einarsson G, Tunney MM, Elborn JS, **Mall MA***, Dalpke AH*. Comparison of microbiomes from different niches of upper and lower airways in children and adolescents with cystic fibrosis. *PLoS ONE* 2015;10:e0116029. (*****equal contribution)

Hector A, Schafer H, Poschel S, Fischer A, Fritzsching B, Ralhan A, Carevic M, Oz H, Zundel S, Hogardt M, Bakele M, Rieber N, Riethmueller J, Graepler-Mainka U, Stahl M, Bender A, Frick JS, **Mall M**, Hartl D. Regulatory T-cell impairment in cystic fibrosis patients with chronic pseudomonas infection. *Am J Respir Crit Care Med* 2015;191:914-923.

Oglesby IK, Agrawal R, **Mall MA**, McElvaney NG, Greene CM. miRNA-221 is elevated in cystic fibrosis airway epithelial cells and regulates expression of ATF6. *Mol Cell Pediatr* 2015;2:1-0012.

Katsirntaki K, Mauritz C, Olmer R, Schmeckebier S, Sgodda M, Puppe V, Eggenschwiler R, Duerr J, Schubert SC, Schmiedl A, Ochs M, Cantz T, Salwig I, Szibor M, Braun T, Rathert C, Martens A, **Mall MA**, Martin U. Bronchoalveolar sublineage specification of pluripotent stem cells: effect of dexamethasone plus cAMP-elevating agents and keratinocyte growth factor. *Tissue Eng Part A* 2015;21:669-682.

Trojanek JB, Cobos-Correa A, Diemer S, Kormann M, Schubert SC, Zhou-Suckow Z, Agrawal R, Duerr J, Wagner CJ, Schatterny J, Hirtz S, Sommerburg O, Hartl D, Schultz C, Mall MA. Airway mucus obstruction triggers macrophage activation and matrix metalloproteinase 12-dependent emphysema. *Am J Respir Cell Mol Biol* 2014;51:709-720.

Gehrig S, Duerr J, Weitnauer M, Wagner CJ, Graeber SY, Schatterny J, Hirtz S, Belaaouaj A, Dalpke AH, Schultz C, **Mall MA**. Lack of Neutrophil Elastase Reduces Inflammation, Mucus Hypersecretion, and Emphysema, but Not Mucus Obstruction, in Mice with Cystic Fibrosis-like Lung Disease. *Am J Respir Crit Care Med* 2014;189:1082-1092.

Wielputz MO, Puderbach M, Kopp-Schneider A, Stahl M, Fritzsching E, Sommerburg O, Ley S, Sumkauskaite M, Biederer J, Kauczor HU, Eichinger M, **Mall MA**. Magnetic resonance imaging detects changes in structure and perfusion, and response to therapy in early cystic fibrosis lung disease. *Am J Respir Crit Care Med* 2014;189:956-965.

Stahl M, Joachim C, Blessing K, Hammerling S, Sommerburg O, Latzin P, **Mall MA**. Multiple breath washout is feasible in the clinical setting and detects abnormal lung function in infants and young children with cystic fibrosis. *Respiration* 2014;87:357-363.

Geiser M, Wigge C, Conrad ML, Eigeldinger-Berthou S, Kunzi L, Garn H, Renz H, **Mall MA**. Nanoparticle uptake by airway phagocytes after fungal spore challenge in murine allergic asthma and chronic bronchitis. *BMC Pulm Med* 2014;14:116.

Jaecklin T, Duerr J, Huang H, Rafii M, Bear CE, Ratjen F, Pencharz P, Kavanagh BP, **Mall MA**, Grasemann H. Lung arginase expression and activity is increased in cystic fibrosis mouse models. *J Appl Physiol* 2014;117:284-288.

Sommerburg O, Krulisova V, Hammermann J, Lindner M, Stahl M, Muckenthaler M, Kohlmueller D, Happich M, Kulozik AE, Votava F, Balascakova M, Skalicka V, Stopsack M, Gahr M, Macek M, Jr., **Mall MA**[*], Hoffmann GF[*]. Comparison of different IRT-PAP protocols to screen newborns for cystic fibrosis in three central European populations. *J Cyst Fibros* 2014;13:15-23. (*****equal contribution)

Bauman G, Puderbach M, Heimann T, Kopp-Schneider A, Fritzsching E, **Mall MA**, Eichinger M. Validation of Fourier decomposition MRI with dynamic contrast-enhanced MRI using visual and automated scoring of pulmonary perfusion in young cystic fibrosis patients. *Eur J Radiol* 2013;82:2371-2377.

Wielputz MO, Weinheimer O, Eichinger M, Wiebel M, Biederer J, Kauczor HU, Heussel CP, **Mall MA**[*], Puderbach M*. Pulmonary emphysema in cystic fibrosis detected by densitometry on chest multidetector computed tomography. *PLOS ONE* 2013;8:e73142. (*****equal contribution)

Graeber SY, Zhou-Suckow Z, Schatterny J, Hirtz S, Boucher RC, **Mall MA**. Hypertonic saline is effective in the prevention and treatment of mucus obstruction, but not airway inflammation, in mice with chronic obstructive lung disease. *Am J Respir Cell Mol Biol* 2013;49:410-417.

Benesova K, Schaefer SM, **Mall MA**, Muckenthaler MU. Computerized image analysis of iron-stained macrophages. *Ann Hematol* 2013;92:1195-1199.

Geiser M, Quaile O, Wenk A, Wigge C, Eigeldinger-Berthou S, Hirn S, Schaffler M, Schleh C, Moller W, **Mall MA**, Kreyling WG. Cellular uptake and localization of inhaled gold nanoparticles in lungs of mice with chronic obstructive pulmonary disease. *Part Fibre Toxicol* 2013;10:19.

Wielputz MO, Eichinger M, Weinheimer O, Ley S, **Mall MA**, Wiebel M, Bischoff A, Kauczor HU, Heussel CP, Puderbach M. Automatic airway analysis on multidetector computed tomography in cystic fibrosis: correlation with pulmonary function testing. *J Thorac Imaging* 2013;28:104-113.

Schmeckebier S, Mauritz C, Katsirntaki K, Sgodda M, Puppe V, Duerr J, Schubert SC, Schmiedl A, Lin Q, Palecek J, Draeger G, Ochs M, Zenke M, Cantz T, **Mall MA**, Martin U. Keratinocyte growth factor and dexamethasone plus elevated cAMP levels synergistically support pluripotent stem cell differentiation into alveolar epithelial type II cells. *Tissue Eng Part A* 2013;19:938-951.

Bingle L, Wilson K, Musa M, Araujo B, Rassl D, Wallace WA, Leclair EE, Mauad T, Zhou Z, **Mall MA**, Bingle CD. BPIFB1 (LPLUNC1) is upregulated in cystic fibrosis lung disease. *Histochem Cell Biol* 2012;138:749-758.

Anagnostopoulou P, Riederer B, Duerr J, Michel S, Binia A, Agrawal R, Liu X, Kalitzki K, Xiao F, Chen M, Schatterny J, Hartmann D, Thum T, Kabesch M, Soleimani M, Seidler U, **Mall MA**. SLC26A9-mediated chloride secretion prevents mucus obstruction in airway inflammation. *J Clin Invest* 2012;122:3629-3634.

Johannesson B, Hirtz S, Schatterny J, Schultz C, **Mall MA**. CFTR regulates early pathogenesis of chronic obstructive lung disease in βENaC-overexpressing mice. *PLoS ONE* 2012;7:e44059.

Benesova K, Vujic SM, Schaefer SM, Stolte J, Baehr-Ivacevic T, Waldow K, Zhou Z, Klingmueller U, Benes V, **Mall MA***, Muckenthaler MU*. Hfe deficiency impairs pulmonary neutrophil recruitment in response to inflammation. *PLoS ONE* 2012;7:e39363. (*****equal contribution)

Gehrig S, **Mall MA***, Schultz C*. Spatially resolved monitoring of neutrophil elastase activity with ratiometric fluorescent reporters. *Angew Chem Int Ed Engl* 2012;51:6258-6261. (*****equal contribution)

Mundhenk L, Johannesson B, Anagnostopoulou P, Braun J, Bothe MK, Schultz C, **Mall MA***, Gruber AD*. mCLCA3 Does Not Contribute to Calcium-Activated Chloride Conductance in Murine Airways. *Am J Respir Cell Mol Biol* 2012;47:87-93. (*****equal contribution)

Hentschel J, Riesener G, Nelle H, Stuhrmann M, Schoner A, Sommerburg O, Fritzsching E, **Mall MA**, von Eggeling F, Mainz JG. Homozygous CFTR mutation M348K in a boy with respiratory symptoms and failure to thrive. Disease-causing mutation or benign alteration? *Eur J Pediatr* 2012;171:1039-1046.

Eichinger M, Optazaite DE, Kopp-Schneider A, Hintze C, Biederer J, Niemann A, **Mall MA**, Wielputz MO, Kauczor HU, Puderbach M. Morphologic and functional scoring of cystic fibrosis lung disease using MRI. *Eur J Radiol* 2012;81:1321-1329.

Wielputz MO, Eichinger M, Zhou Z, Leotta K, Hirtz S, Bartling SH, Semmler W, Kauczor HU, Puderbach M, **Mall MA**. In vivo monitoring of cystic fibrosis-like lung disease in mice by volumetric computed tomography. *Eur Respir J* 2011;38:1060-1070.

Roth EK, Hirtz S, Duerr J, Wenning D, Eichler I, Seydewitz HH, Amaral MD, **Mall MA**. The K^+ channel opener 1-EBIO potentiates residual function of mutant CFTR in rectal biopsies from cystic fibrosis patients. *PLoS ONE* 2011;6:e24445.

Hector A, Kormann MS, Mack I, Latzin P, Casaulta C, Kieninger E, Zhou Z, Yildirim AO, Bohla A, Rieber N, Kappler M, Koller B, Eber E, Eickmeier O, Zielen S, Eickelberg O, Griese M, **Mall MA**, Hartl D. The chitinase-like protein YKL-40 modulates cystic fibrosis lung disease. *PLoS ONE* 2011;6:e24399.

Bonora M, Riffault L, Marie S, **Mall M**, Clement A, Tabary O. Morphological analysis of the trachea and pattern of breathing in βENaC-Tg mice. *Respir Physiol Neurobiol* 2011;178:346-348.

Duerr J, Gruner M, Schubert SC, Haberkorn U, Bujard H, **Mall MA**. Use of a new-generation reverse tetracycline transactivator system for quantitative control of conditional gene expression in the murine lung. *Am J Respir Cell Mol Biol* 2011;44:244-254.

Anagnostopoulou P, Dai L, Schatterny J, Hirtz S, Duerr J, **Mall MA**. Allergic airway inflammation induces a pro-secretory epithelial ion transport phenotype in mice. *Eur Respir J* 2010;36:1436-1447.

Sommerburg O, Lindner M, Muckenthaler M, Kohlmueller D, Leible S, Feneberg R, Kulozik AE, **Mall MA***, Hoffmann GF*. Initial evaluation of a biochemical cystic fibrosis newborn screening by sequential analysis of immunoreactive trypsinogen and pancreatitis-associated protein (IRT/PAP) as a strategy that does not involve DNA testing in a Northern European population. *J Inherit Metab Dis* 2010;33:S263-S271. (*equal contribution)

Mall MA, Button B, Johannesson B, Zhou Z, Livraghi A, Caldwell RA, Schubert SC, Schultz C, O'Neal WK, Pradervand S, Hummler E, Rossier BC, Grubb BR, Boucher RC. Airway surface liquid volume regulation determines different airway phenotypes in liddle compared with βENaC-overexpressing mice. *J Biol Chem* 2010;285:26945-26955.

Cobos-Correa A, **Mall MA**, Schultz C. Reporters to monitor cellular MMP12 activity. *Proc of SPIE* 2010;7576:1-10.

Cobos-Correa A, Trojanek JB, Diemer S, **Mall MA***, Schultz C*. Membrane-bound FRET probe visualizes MMP12 activity in pulmonary inflammation. *Nat Chem Biol* 2009;5:628-630. (*equal contribution)

Livraghi A, Grubb BR, Hudson EJ, Wilkinson KJ, Sheehan JK, **Mall MA**, O'Neal WK, Boucher RC, Randell SH. Airway and lung pathology due to mucosal surface dehydration in β-epithelial Na+ channel-overexpressing mice: role of TNF-α and IL-4Rα signaling, influence of neonatal development, and limited efficacy of glucocorticoid treatment. *J Immunol* 2009;182:4357-4367.

Zhou Z, Treis D, Schubert S, Harm M, Schatterny J, Hirtz S, Duerr J, Boucher RC, **Mall MA**. Preventive but not late amiloride therapy reduces morbidity and mortality of lung disease in βENaC-overexpressing mice. *Am J Respir Crit Care Med* 2008;178:1245-1256.

Mall MA, Harkema JR, Trojanek JB, Treis D, Livraghi A, Schubert S, Zhou Z, Kreda SM, Tilley SL, Hudson EJ, O'Neal WK, Boucher RC. Development of chronic bronchitis and emphysema in βENaC-overexpressing mice. *Am J Respir Crit Care Med* 2008;177: 730-42.

Livraghi A, **Mall M**, Paradiso AM, Boucher RC, Pedrosa Ribeiro CM. Modelling Dysregulated Na^+ Absorption in Airway Epithelial Cells with Mucosal Nystatin Treatment. *Am J Respir Cell Mol Biol* 2008;38:423-34.

Kreda SM, **Mall M**, Mengos A, Rochelle L, Yankaskas J, Riordan JR, Boucher RC. Characterization of wild-type and deltaF508 cystic fibrosis transmembrane regulator in human respiratory epithelia. *Mol Biol Cell* 2005;16:2154-2167.

Kunzelmann K, Sun J, Markovich D, Konig J, Murle B, **Mall M**, Schreiber R. Control of ion transport in mammalian airways by protease activated receptors type 2 (PAR-2). *FASEB J* 2005;19:969-976.

Mall M, Grubb BR, Harkema JR, O'Neal WK, Boucher RC. Increased airway epithelial Na^+ absorption produces cystic fibrosis-like lung disease in mice. *Nat Med* 2004;10:487-493.

Hirtz S, Gonska T, Seydewitz HH, Thomas J, Greiner P, Kuehr J, Brandis M, Eichler I, Rocha H, Lopes AI, Barreto C, Ramalho A, Amaral MD, Kunzelmann K, **Mall M.** CFTR Cl- channel function in native human colon correlates with the genotype and phenotype in cystic fibrosis. *Gastroenterology* 2004;127:1085-1095.

Mall M, Kreda SM, Mengos A, Jensen TJ, Hirtz S, Seydewitz HH, Yankaskas J, Kunzelmann K, Riordan JR, Boucher RC. The DeltaF508 mutation results in loss of CFTR function and mature protein in native human colon. *Gastroenterology* 2004;126:32-41.

Mall M, Gonska T, Thomas J, Schreiber R, Seydewitz HH, Kuehr J, Brandis M, Kunzelmann K. Modulation of Ca^{2+}-activated Cl^- secretion by basolateral K^+ channels in human normal and cystic fibrosis airway epithelia. *Pediatr Res* 2003;53:608-618.

Ramalho AS, Beck S, Penque D, Gonska T, Seydewitz HH, **Mall M**, Amaral MD. Transcript analysis of the cystic fibrosis splicing mutation 1525-1G>A shows use of multiple alternative splicing sites and suggests a putative role of exonic splicing enhancers. *J Med Genet* 2003;40:e88.

Mall M, Gonska T, Thomas J, Hirtz S, Schreiber R, Kunzelmann K. Activation of ion secretion via proteinase-activated receptor-2 in human colon. *Am J Physiol Gastrointest Liver Physiol* 2002;282:G200-G210.

Konig J, Schreiber R, **Mall M**, Kunzelmann K. No evidence for inhibition of ENaC through CFTR-mediated release of ATP. *Biochim Biophys Acta* 2002;1565:17-28.

Konig J, Schreiber R, Voelcker T, **Mall M**, Kunzelmann K. The cystic fibrosis transmembrane conductance regulator (CFTR) inhibits ENaC through an increase in the intracellular Cl- concentration. *EMBO Rep* 2001;2:1047-1051.

Mall M, Wissner A, Seydewitz HH, Kuehr J, Brandis M, Greger R, Kunzelmann K. Defective cholinergic Cl^- secretion and detection of K^+ secretion in rectal biopsies from cystic fibrosis patients. *Am J Physiol Gastrointest Liver Physiol* 2000;278:G617-G624.

Seydewitz HH, **Mall M**, Kuehr J. A novel missense mutation, S1159F, in exon 19 of the CFTR gene. *Hum Mutat* 2000;15:390.

Mall M, Wissner A, Schreiber R, Kuehr J, Seydewitz HH, Brandis M, Greger R, Kunzelmann K. Role of K_VLQT1 in cAMP-mediated Cl^- secretion in human airway epithelia. *Am J Respir Cell Molec Biol* 2000;23:283-289.

Mall M, Wissner A, Seydewitz HH, Hübner M, Kuehr J, Brandis M, Greger R, Kunzelmann K. Effect of genistein on native epithelial tissue from normal individuals and CF patients and on ion channels expressed in Xenopus oocytes. *Br J Pharmacol* 2000;130:1884-1892.

Seydewitz HH, Gonska T, **Mall M**, Kuehr J. A novel frameshift mutation, 1870delG, in exon 12 of the CFTR gene. *Hum Mutat*, 2000;16:277.

Mall M, Wissner A, Gonska T, Calenborn D, Kuehr J, Brandis M, Kunzelmann K. Inhibition of amiloride-sensitive epithelial Na^+ absorption by extracellular nucleotides in human normal and cystic fibrosis airways. *Am J Respir Cell Mol Biol* 2000;23:755-761.

Hopf A, Schreiber R, **Mall M**, Greger R, Kunzelmann K. Cystic fibrosis transmembrane conductance regulator inhibits epithelial Na^+ channels carrying Liddle's syndrome mutations. *J Biol Chem* 1999;274:13894-13899.

Schreiber R, Hopf A, **Mall M**, Greger R, Kunzelmann K. The first nucleotide binding domain of the cystic fibrosis transmembrane conductance regulator is important for inhibition of the epithelial Na^+ channel. *Proc Natl Acad Sci USA* 1999;96:5310-5315.

Mall M, Bleich M, Kuehr J, Brandis M, Greger R, Kunzelmann K. CFTR-mediated inhibition of epithelial Na^+ conductance in human colon is defective in cystic fibrosis. *Am J Physiol Gastrointest Liver Physiol* 1999;277:G709-G716.

Mall M, Bleich M, Greger R, Schreiber R, Kunzelmann K. The amiloride inhibitable Na^+ conductance is reduced by CFTR in normal but not in cystic fibrosis airways. *J Clin Invest* 1998;102:15-21.

Mall M, Bleich M, Greger R, Schürlein M, Kühr J, Seydewitz HH, Brandis M, Kunzelmann K. Cholinergic ion secretion in human colon requires co-activation by cAMP. *Am J Physiol* 1998;275:G1274-G1281.

Kunzelmann K, **Mall M**, Briel M, Hipper A, Nitschke R, Ricken S, Greger R. The cystic fibrosis transmembrane conductance regulator attenuates the endogenous Ca^{2+} activated Cl^- conductance in *Xenopus* ooyctes. *Eur J Physiol -Pflügers Arch* 1997;434:178-18.

Mall M, Hipper A, Greger R, Kunzelmann K. Wild type but not F508 CFTR inhibits Na^+ conductance when coexpressed in *Xenopus* oocytes. *FEBS Lett* 1996;381:47-52.

Mall M, Kunzelmann K, Hipper A, Busch AE, Greger R. cAMP stimulation of CFTR expressing *xenopus* oocytes activates a chromanol inhibitable K^+ conductance. *Eur J Physiol -Pflügers Arch* 1996;432:516-522.

Hipper A, **Mall M**, Greger R, Kunzelmann K. Mutations in the putative pore forming domain of CFTR do not change anion selectivity of the cAMP activated Cl^- conductance. *FEBS Lett* 1995;374:312-316.

Übersichtsartikel und Editorials

Graeber SY, Vitzthum C, **Mall MA**. Potential of Intestinal Current Measurement for Personalized Treatment of Patients with Cystic Fibrosis. *J Pers Med* 2021;11.

McKelvey MC, Brown R, Ryan S, **Mall MA**, Weldon S, Taggart CC. Proteases, Mucus, and Mucosal Immunity in Chronic Lung Disease. *Int J Mol Sci* 2021;22.

Stahl M, Steinke E, **Mall MA**. Quantification of Phenotypic Variability of Lung Disease in Children with Cystic Fibrosis. *Genes (Basel)* 2021;12.

Mall MA. ENaC inhibition in cystic fibrosis: potential role in the new era of CFTR modulator therapies. *Eur Respir J* 2020;56:2000946.

Leutz-Schmidt P, Eichinger M, Sommerburg O, Stahl M, Triphan SMF, Gehlen S, Kauczor HU, Puderbach MU, **Mall MA**, Wielputz MO. Magnetic resonance imaging of the lungs in cystic fibrosis (Magnetresonanztomographie der Lunge bei Mukoviszidose). *Radiologe* 2020;60:813-822.

Bischoff A, Weinheimer O, Eichinger M, Stahl M, Sommerburg O, Kauczor HU, Mall MA, Wielpütz MO. Cystic fibrosis and computed tomography of the lungs (Computertomographie der Lunge bei Mukoviszidose). *Radiologe* 2020;60:791-801.

Mall MA*, Mayer-Hamblett N*, Rowe SM*. Cystic fibrosis: Emergence of highly effective targeted therapeutics and potential clinical implications. *Am J Respir Crit Care Med* 2020;201:1193-1208. (*equal contribution).

Taggart CC, Weldon S, **Mall MA**. At the forefront of cystic fibrosis basic science research: 16th ECFS basic science conference. *J Cyst Fibros* 2020;19:169-170.

Giacalone VD, Margaroli C, **Mall MA**, Tirouvanziam R. Neutrophil adaptations upon recruitment to the lung: New concepts and implications for homeostasis and disease. *Int J Mol Sci* 2020;21:e851.

McKelvey MC, Weldon S, McAuley DF, **Mall MA***, Taggart CC*. Targeting proteases in cystic fibrosis lung disease: Paradigms, progress, and potential. *Am J Respir Crit Care Med* 2020;201(2):141–147. (*equal contribution).

Bell SC, **Mall MA**, Gutierrez H, Macek M, Madge S, Davies JC, Burgel PR, Tullis E, Castanos C, Castellani C, Byrnes CA, Cathcart F, Chotirmall SH, Cosgriff R, Eichler I, Fajac I, Goss CH, Drevinek P, Farrell PM, Gravelle AM, Havermans T, Mayer-Hamblett N, Kashirskaya N, Kerem E, Mathew JL, McKone EF, Naehrlich L, Nasr SZ, Oates GR, O'Neill C, Pypops U, Raraigh KS, Rowe SM, Southern KW, Sivam S, Stephenson AL, Zampoli M, Ratjen F. The future of cystic fibrosis care: A global perspective. *Lancet Respir Med* 2020;8:65–124.

Leutz-Schmidt P, Eichinger M, Stahl M, Sommerburg O, Biederer J, Kauczor HU, Puderbach MU, **Mall MA**, Wielputz MO. Ten years of chest MRI for patients with cystic fibrosis: Translation from the bench to clinical routine. *Radiologe* 2019;59:10-20.

Balazs A, **Mall MA**. Mucus obstruction and inflammation in early cystic fibrosis lung disease: Emerging role of the IL-1 signaling pathway. *Pediatr Pulmonol* 2019; 54 Suppl 3: S5-S12.

Stahl M, Joachim C, Wielputz MO, **Mall MA**. Authors' response: Letter to the editor 'comparison of lung clearance index determined by washout of N2 and SF6 in infants and preschool children with cystic fibrosis'. *J Cyst Fibros* 2019; 18: e28-e29.

Mall MA, Danahay H, Boucher RC. Emerging Concepts and Therapies for Mucoobstructive Lung Disease. *Ann Am Thorac Soc* 2018;15:S216-S226.

Tümmler B, **Mall MA**. Nicht nur Mukoviszidose: Diagnostik und Therapie CFTR-assoziierter Störungen. *Monatsschr Kinderheilkd* 2018;166: Zertifizierte Fortbildung

Balazs A, **Mall MA**. Role of the SLC26A9 Chloride Channel as Disease Modifier and Potential Therapeutic Target in Cystic Fibrosis. *Front Pharmacol* 2018;9:1112.

Gentzsch M, **Mall MA**. Ion Channel Modulators in Cystic Fibrosis. *Chest* 2018;154:383-393.

Mall MA, Hwang TC, Braakman I. Cystic fibrosis research topics featured at the 14th ECFS Basic Science Conference: Chairman's summary. *J Cyst Fibros* 2018;17:S1-S4.

Balazs A, **Mall MA**. Mucopurulent Triggering of the Airway Epithelium. Implications in Health and Cystic Fibrosis. *Am J Respir Crit Care Med* 2018;197:418-420.

Stahl M, Wielputz MO, Kauczor HU, **Mall MA**. Reply to Verbanck and Vanderhelst: The Respective Roles of Lung Clearance Index and Magnetic Resonance Imaging in the Clinical Management of Patients with Cystic Fibrosis. *Am J Respir Crit Care Med* 2018;197:410-411.

Li H, Salomon JJ, Sheppard DN, **Mall MA***, Galietta LJ*. Bypassing CFTR dysfunction in cystic fibrosis with alternative pathways for anion transport. *Curr Opin Pharmacol* 2017;34:91-97. (*****equal contribution)

Taggart C, **Mall MA**, Lalmanach G, Cataldo D, Ludwig A, Janciauskiene S, Heath N, Meiners S, Overall CM, Schultz C, Turk B, Borensztajn KS. Protean proteases: at the cutting edge of lung diseases. *Eur Respir J* 2017;49:1501200.

Montgomery ST, **Mall MA***, Kicic A*, Stick SM*. Hypoxia and sterile inflammation in cystic fibrosis airways: mechanisms and potential therapies. *Eur Respir J* 2017;49:1600903. (*equal contribution)

Zhou-Suckow Z, Duerr J, Hagner M, Agrawal R, **Mall MA**. Airway mucus, inflammation and remodeling: emerging links in the pathogenesis of chronic lung diseases. *Cell Tissue Res* 2017;367:537-550.

Elborn JS, Davies J, **Mall MA**, Flume PA, Plant B. Current strategies for the long-term assessment, monitoring, and management of cystic fibrosis patients treated with CFTR modulator therapy. *J Cyst Fibros* 2017;16:163-164.

Wielpütz MO, **Mall MA**. MRI accelerating progress in functional assessment of cystic fibrosis lung disease. *J Cyst Fibros* 2017;16:165-167.

Sommerburg O, Stahl M, Hämmerling S, Lawrenz B, **Mall MA**, Hoffmann GF. Einführung des Neugeborenen-Screenings auf Mukoviszidose in Deutschland. *Pädiatrische Praxis* 2017;87:569-580.

Sommerburg O, Lawrenz B, **Mall MA**, Hoffmann GF. Das Mukoviszidosescreening wird in Deutschland eingeführt: Was müssen die Pädiater in der Praxis wissen? *Monatsschr Kinderheilkd* 2017;165:49-54.

Rickert-Zacharias V, Schultz C, **Mall MA**. A protease inhibitor tackles epithelial sodium channels in cystic fibrosis. *Am J Respir Crit Care Med* 2016;194:650-652.

Mall MA, Stahl M, Graeber SY, Sommerburg O, Kauczor HU, Wielputz MO. Early detection and sensitive monitoring of CF lung disease: Prospects of improved and safer imaging. *Pediatr Pulmonol* 2016;51:S49-S60.

Wielputz MO, Eichinger M, Biederer J, Wege S, Stahl M, Sommerburg O, **Mall MA**, Kauczor HU, Puderbach M. Imaging of cystic fibrosis lung disease and clinical interpretation. *Rofo* 2016;188:834-845.

Wagner CJ, Schultz C, **Mall MA**. Neutrophil elastase and matrix metalloproteinase 12 in cystic fibrosis lung disease. *Mol Cell Pediatr* 2016;3:25.

Mall MA. Unplugging mucus in cystic fibrosis and chronic obstructive pulmonary disease. *Ann Am Thorac Soc* 2016;13 Suppl 2:S177-85.

Mall MA. Mukoviszidose – neue Therapieansätze. *Frankfurter Allgemeine Zeitung* Verlagsspezial Lungengesundheit 2015:V2.

Mall MA, Galietta LJ. Targeting ion channels in cystic fibrosis. *J Cyst Fibros* 2015;14:561-570.

Wielputz MO, **Mall MA**. Imaging modalities in cystic fibrosis: emerging role of MRI. *Curr Opin Pulm Med* 2015;21:609-616.

Sommerburg O, Schenk JP, **Mall MA**. [Lung diseases in children]. *Radiologe* 2015;55:545-553.

Verkman AS, Edelman A, Amaral M, **Mall MA**, Beekman JM, Meiners T, Galietta LJ, Bear CE. Finding new drugs to enhance anion secretion in cystic fibrosis: Toward suitable systems for better drug screening. Report on the pre-conference meeting to the 12th ECFS Basic Science Conference, Albufeira, 25-28 March 2015. *J Cyst Fibros* 2015;14:700-705.

Palaniyar N, **Mall MA**, Taube C, Worgall S, Grasemann H. New Developments in Cystic Fibrosis Airway Inflammation. *Mediators Inflamm* 2015;2015:769425.

Mack I, Hector A, Ballbach M, Kohlhaufl J, Fuchs KJ, Weber A, **Mall MA**, Hartl D. The role of chitin, chitinases, and chitinase-like proteins in pediatric lung diseases. *Mol Cell Pediatr* 2015;2:3-0014.

Mall MA, Sheppard DN. Chronic ivacaftor treatment: getting F508del-CFTR into more trouble? *J Cyst Fibros* 2014;13:605-607.

Grether M, Eickelberg O, **Mall MA**, Rabe KF, Welte T, Seeger W. New metrics for translational research. *Lancet Respir Med* 2014;2:e13-e14.

Mall MA, Hartl D. CFTR: cystic fibrosis and beyond. *Eur Respir J* 2014;44:1042-1054.

Mall MA, Schultz C. A new player in the game: epithelial cathepsin S in early cystic fibrosis lung disease. *Am J Respir Crit Care Med* 2014;190:126-127.

Mall MA, Graeber SY, Stahl M, Zhou-Suckow Z. Early cystic fibrosis lung disease: Role of airway surface dehydration and lessons from preventive rehydration therapies in mice. *Int J Biochem Cell Biol* 2014;52:174-179.

Beekman JM, Sermet-Gaudelus I, De Boeck K, Gonska T, Derichs N, **Mall MA**, Mehta A, Martin U, Drumm M, Amaral MD. CFTR functional measurements in human models for diagnosis, prognosis and personalized therapy. *J Cyst Fibros* 2014;13:363-372.

Hu HY, Gehrig S, Reither G, Subramanian D, **Mall MA**, Plettenburg O, Schultz C. FRET-based and other fluorescent proteinase probes. *Biotechnol J* 2014;9:266-281.

Hartl D, Gaggar A, Bruscia E, Hector A, Marcos V, Jung A, Greene C, McElvaney G, **Mall M**, Doring G. Innate immunity in cystic fibrosis lung disease. *J Cyst Fibros* 2012;11:363-382.

Seeger W, Welte T, Eickelberg O, **Mall M**, Rabe KF, Keller B, Winkler S, Koller U. The German Centre for Lung Research - Translational Research for the Prevention, Diagnosis and Treatment of Respiratory Diseases. *Pneumologie* 2012;66:464-469.

Moog U, Okun PM, Bettendorf M, Gorenflo M, Kölker S, Krämer A, Lorenz HM, **Mall MA**, Muckenthaler MU, Schaefer FS, Schönland SO, Hoffmann GF. Zentrum für Seltene Erkrankungen Universitätsmedizin Heidelberg. *Medizinische Genetik* 2011;4:493-504.

Gaggar A, Hector A, Bratcher PE, **Mall MA**, Griese M, Hartl D. The role of matrix metalloproteinases in cystic fibrosis lung disease. *Eur Respir J* 2011;38:721-727.

Zhou Z, Duerr J, Johannesson B, Schubert SC, Treis D, Harm M, Graeber SY, Dalpke A, Schultz C, **Mall MA**. The ENaC-overexpressing mouse as a model of cystic fibrosis lung disease. *J Cyst Fibros* 2011;10 Suppl 2:S172-S182.

Becq F, **Mall MA**, Sheppard DN, Conese M, Zegarra-Moran O. Pharmacological therapy for cystic fibrosis: from bench to bedside. *J Cyst Fibros* 2011;10 Suppl 2:S129-S145.

Sheppard DN, Liu J, Zhou Z, **Mall MA**. Treatment of cystic fibrosis lung disease - the development of drugs targeting cystic fibrosis transmembrane conductance regulator dysfunction. *European Respiratory Disease* 2011;7:12-17.

Mall MA. Role of the amiloride-sensitive epithelial Na^+ channel in the pathogenesis and as a therapeutic target for cystic fibrosis lung disease. *Exp Physiol* 2009;94:171-174.

Mall MA. Role of cilia, mucus and airway surface liqiud in mucociliary dysfunction: lessons from mouse models. *J Aerosol Med Pulm Drug Deliv* 2008;21:13-24.

Mall M. Hypertones Kochsalz: Therapie der Lungenerkrankung bei Mukoviszidose - Mechanismen und Langzeitwirkung. *Monatsschr Kinderheilkd* 2006;154:907-909.

Mall M, Kunzelmann K. Correction of the CF defect by curcumin: hypes and disappointments. *Bioessays* 2005;27:9-13.

Mall M, Hirtz S, Gonska T, Kunzelmann K. Assessment of CFTR function in rectal biopsies for the diagnosis of cystic fibrosis. *J Cyst Fibros* 2004;3 Suppl 2:165-169.

Kunzelmann K, **Mall M**. Pharmacotherapy of the ion transport defect in cystic fibrosis: role of purinergic receptor agonists and other potential therapeutics. *Am J Respir Med* 2003;2:299-309.

Kunzelmann K, **Mall M**. Electrolyte transport in the mammalian colon: mechanisms and implications for disease. *Physiol Rev* 2002;82:245-289.

Kunzelmann K, Schreiber R, Konig J, **Mall M**. Ion transport induced by proteinase-activated receptors (PAR2) in colon and airways. *Cell Biochem Biophys* 2002;36:209-214.

Kunzelmann K, **Mall M**. Pharmacotherapy of the ion transport defect in cystic fibrosis. *Clin Exp Pharmacol Physiol* 2001;28:857-867.

Greger R, Schreiber R, **Mall M**, Wissner A, Hopf A, Briel M, Bleich M, Warth R, Kunzelmann K. Cystic fibrosis and CFTR. *Eur J Physiol - Pflügers Arch* 2001;443:S3-S7.

Ballmann M, Griese M, Hebestreit A, Hofmann T, Kersting U, Lindemann H, **Mall M**, Schüler D, Reinhardt D. Klinischer Stellenwert der modernen elektrophysiologischen Diagnostik bei Mukoviszidose (Zystischer Fibrose). Monatsschr Kinderheilkd 2001;149:258-264.

Kunzelmann K, Schreiber R, Nitschke R, **Mall M**. Control of the epithelial Na^+ conductance by the cystic fibrosis transmembrane regulator. *Eur J Physiol -Pflügers Arch* 2000;440:193-201.

Greger R, Bleich M, Leipziger J, Ecke D, **Mall M**, Kunzelmann K. Regulation of ion transport in colonic crypts. *News Physiol Sci* 1997;12:62-66.

Greger R, **Mall M**, Bleich M, Ecke D, Warth R, Riedemann N, Kunzelmann K. Regulation of epithelial ion channels by the cystic fibrosis transmembrane conductance regulator (CFTR). *J Mol Med* 1996;74:527-534.

Buchkapitel

Mall MA, Sommerburg O. Mukoviszidose. In: Kreuter M, Costabel U, Herth F, and Kirsten D, eds. Seltene Lungenerkrankungen. Berlin Heidelberg: Springer Verlag, 2016:303-311.

Mall MA, Graeber SY. Biophysikalische Grundlagen der Anwendung. In: Möller A, ed. Hypertone Salzlösung bei Atemwegserkrankungen - Grundlagen und praktische Anwendung. Bremen: UNI-MED, 2015:11-21.

Mall MA, Graeber SY. Bioassays to assess CFTR function in humans: intestinal current measurements. In: Tümmler B, ed. Mutation-specific therapies in cystic fibrosis - Current status and prospects. Bremen: UNI-MED, 2014:40-44.

Mall MA, Aumann I., Prenzler A., Graf v.d.Schulenburg J.-M. Mukoviszidose (Zystische Fibrose, CF). In: Gillissen A. and Welte T, eds. Weißbuch Lunge 2014: Die Lunge und ihre Erkrankungen: Zur Lage und Zukunft der Pneumologie in Deutschland. Herne: Frischtexte Verlag, 2014:137-142.

Mall MA, Boucher RC. Pathophysiology of cystic fibrosis lung disease. In: Mall MA and Elborn JS, eds. Cystic fibrosis. Sheffield: European Respiratory Society, 2014:1-13.

Mall MA. CFTR-Funktionsdiagnostik. In: von Mutius E, Gappa M, Eber E, and Frey U, eds. Pädiatrische Pneumologie. 3. Auflage ed. Berlin Heidelberg: Springer Verlag, 2014:307-312.

Mall MA, Reinhardt D. Erkrankungen der Atemwegsorgane. In: Koletzko B, ed. Kinder- und Jugendmedizin. 14. Auflage ed. Berlin Heidelberg: Springer Verlag, 2013:365-407.

Sommerburg O, **Mall M**. Molekulare Grundlage der Sekretretention und Mukostase. In: Hirche TO and Wagner TOF, eds. Update Mukoviszidose. Stuttgart, New York: Georg Thieme Verlag, 2010:10-11.

Kunzelmann K, **Mall M**. Cystic fibrosis. In: Lang F, ed. Encyclopedia of molecular mechanisms of disease. Berlin Heidelberg: Springer Verlag, 2009:482-483

Sommerburg O, **Mall M**. Pathophysiologie der Lungenerkrankung bei CF - aktuelle Hypothesen. In: Hirche TO and Wagner TOF, eds. Update Mukoviszidose. Stuttgart, New York: Georg Thieme Verlag, 2009:7-9

Mall M, Boucher RC. Pathogenesis of pulmonary disease in cystic fibrosis. In: Bush A, Alton E, Davies J, Griesenbach U, and Jaffe A, eds. Cystic fibrosis in the 21st century. *Prog Respir Res*. Basel: Karger, 2006:116-121.

Kunzelmann K, Bachhuber T, Adam G, Voelker T, Muerle B, **Mall M**, Schreiber R. Role of CFTR and other ion channels in cystic fibrosis. In: Schultz C, ed. Defects of secretion in cystic fibrosis. *Adv Exp Med Biol.* New York: Springer, 2005;558:23-41.

Kunzelmann K, Sun J, Markovich D, König J, Mürle B, Voelker T, **Mall M**, Schreiber R. Inhibition of epithelial Na^+ channels by CFTR and purinergic agonists: mechanisms and significance for CF. In: Schulz I and Isenberg G, eds. Nova Acta Leopoldina: Epithelial transport of ions in health and disease. 89 ed. Halle (Saale): Leopoldina, 2004:177-182.

Monographien und Sonderausgaben

Cystic Fibrosis Research Topics Featured at the 14th ECFS Basic Science Conference, Albufeira, Portugal, 29 March to 1 April, 2017. Edited by **Marcus A. Mall**, Tzyh-Chang Hwang, Ineke Braakman. *J Cyst Fibros* 2018;Volume 17, Issue 2, Supplement, S1-S60

Cystic fibrosis. Edited by **Mall, M. A**. and Elborn, J. S. *ERS Monograph*. Sheffield: European Respiratory Society, 1-319. 2014.

Fallberichte

Hämmerling S, Becker S, **Mall MA**. Rare cause for hemoptysis in an adolescent: Bronchial capillary hemangioma. *Pediatr Pulmonol* 2017;52:E40-E42.

Hämmerling S, Wühl E, Grulich-Henn J, Sommerburg O, **Mall M**, Federspil PA. Stridor bei zwei Jugendlichen. *Pädiatrische Praxis* 2016;85:415-422.

Mall M, Sutor AH. Kontinuierliche postoperative Infusion von Faktor VIII bei Hämophilie A. *Pädiatrische Praxis*, 55:433-438, 1998.