Jane C Davies
Curriculum Vitae

Professional address Dept of CF & Chronic Lung Infection , National Heart & Lung Institute, Imperial College London, Emmanuel Kaye Building, Manresa Rd, London SW3 6LR

E-mail j.c.davies@imperial.ac.uk **GMC** 3261765

Current Posts

Professor of Paediatric Respirology & Experimental Medicine, NHLI, Imperial College London (2013 onwards)

Honorary Consultant in Paediatric Respiratory Medicine
Royal Brompton & Harefield NHS Foundation Trust, London (1999 onwards)

Qualifications

1987	MB ChB, University of Dundee
1991	MRCP (Paeds), London
1996	MRCPCH London
1998	MD (Hons) University of Dundee

Previous Posts

2009 – 2013	Reader in Gene Therapy, Imperial College London
1999 – 2009	Senior Lecturer in Gene Therapy, Imperial College London
1998 – 1999	SpR in Paediatric Immunology and Infectious Disease Great Ormond Street Hospital, London
1997 – 1998	SpR in Paediatric Respiratory Medicine and Intensive Care Great Ormond Street Hospital, London
1994 – 1997	Research Fellow in Paediatric Cystic Fibrosis Royal Brompton & Harefield NHS Foundation Trust, London
1993 – 1994	Registrar in Paediatric Respiratory Medicine & Intensive Care Royal Brompton & Harefield NHS Foundation Trust, London
1992 – 1993	Registrar in Paediatrics and Neonatology Hillingdon Hospital, Middlesex
1991 – 1992	Senior House Officer, Infectious Diseases and Immunology; Neurology; Dermatology, Great Ormond Street Hospital, London
1990 – 1991	Senior House Officer, Haematology; Paediatric Surgery; Gastroenterology; Neonatology, Queen Elizabeth Hospital for Children & Homerton Hospital.
1989 – 1990	Senior House Officer in Paediatrics and Special Care, West Middlesex Hospital, London

Areas of Research Interest

Novel therapies and clinical trial design

Outcome measures for CF lung disease, including young children

Pathogenesis in CF:

Pseudomonas aeruginosa: pathogenesis, antimicrobial therapies and non-invasive detection methods

Innate defence, airway remodelling and inflammation

Students supervised[1]/ co-supervised[2]

BSc/ MBBS

Tamsin Lloyd[1] (2000-01; BSc Uni York)
Robert Shaw[1] (2001-02; BSc Uni York)
Arran Titley[1] (2002-03; BSc Uni York)
Louise Collins[1] (2003- 04; BSc Uni York)
Laura Benzonana[1] (2003-04; BSc Uni York)
Martha Truscott[1] (2004-05; BSc Uni York)
Fiona McLean[1] (2005-06; BSc Uni York)
Laura Wright[1] (2006-07; BSc Uni York)
Robert Fordham[1] (2007-08; BSc Uni York)
Sarah Moody[1] (2008-09; BSc Uni York)
Isis Redford[1] (2009-10; BSc Uni York)
Khayam Naderi[1] (2010; MBBS BSc Imperial)
Rajit Khosla[1] (2011; MBBS BSc Imperial)
Valerie Khoo[1] (2015; ICL MBBS BSc Imperial)
Aaniya Ahmed[1] (2015; MBBS summer student)
Helena Lund Palau[1] (2015; MBBS Erasmus Spain)
Laurance Pallant[1] (2016; MBBS BSc Imperial)
Nisal Weerakoon[1] (2016; MBBS BSc Imperial)
Ishani Seth[1] (2016; MBBS summer student)
Abigail Lark[1] (2017; BSc summer student)
Merel Oudraad[1] (2017 MBBS Erasmus Netherlands)
Erica Asperges[1] (2017 MBBS Erasmus Italy)
Dylan Amin[1] (2019 MBBS; intercalated BSc Imperial)
Frouke Terpstra[1] (2019 MBBS Erasmus Netherlands)
Aditya Malkar[1] (2022 MBBS; intercalated BSc)

MSc/ MRes

Yaqi Hu[1] (Genes, Drugs & Stem cells 2016)
Christopher Short (2018 - 2021)
Matt Besley[1] (Genes, Drugs & Stem cells – 2019)
Asma Alharbi[1] (Healthcare – 2019)
Mickael Aoun[1] (Genes, Drugs & Stem cells-2022)
Marina Kusumoto[1] (Biomedical Research- 2022)

Pre-doctoral fellowship

Sian Bentley (2019 – 2021 NIHR)

MD/ MDRes

Tom Hilliard[1] (2002- 04) MD awarded
Hui-Leng Tan[1] (2007-09) MD awarded
Andrew Ives[2] (2007-09)
Sarah Brown[2] (2009-11); MDRes awarded
Andrew Jones[2] (2009- 13) MD awarded
Rebecca Thursfield[1] (2011-13; MDRes awarded)
Katherine Harman[1] (2012-2015; MDRes awarded)
Tom Semple[1] (2016- 2022) Awarded
Isaac Martin[1] (2016 – 2021) Awarded
Laura Gardner (2018 – writing up)
Chris Hine (Uni Birmingham; 2019- writing up)
John King (2020 – ongoing)
Afsoon Sepahzad (2021-ongoing)
Simone Hadjisymeou-Andreou (2022-ongoing)

PhD

Katy Fidler[2] (2001-07 PhD awarded)
Jackie Donovan[1] (2004-12 part-time; PhD awarded)
Gwyneth Davies[1] (2008- 13; PhD awarded)
Chandrika Nair[2] (2010-14; PhD awarded)
Matthew Coates[1] (2011- 2017 part-time; PhD awarded)
Rishi Pabary[1] (2011-14; PhD awarded)
Mike Waller[2] (2012-14; PhD awarded)
Katie Bayfield[1] (2013- 2016; PhD awarded)
Katie Farrant[2] (2013- 2017; PhD awarded)
Bushra Ahmed[2] (2014 – 2017; PhD awarded)
Andrew Turnbull[1] (2014- 2018; PhD awarded)
Natasha Wierre-Gore[2] (2015- writing up)
Emmanuelle Bardin[2] (2015- 2019; PhD awarded)
Loren Cameron[2] (2015- 2019; PhD awarded)
Wynne Smith[1] (2015- writing up; part-time)
Gemma Stanford[2] (2016- writing up; part-time)
Claire Edmondson[1] (2016- 2020; PhD awarded)
Dominic Hughes[1] (2017–2022; PhD awarded)

Ronan Murphy[1] (2017 – 2021; PhD awarded)
Rebecca Dobra[1] (2017- 2022; PhD awarded)
Livia Spiga[2] (2018-2022; submitted)
Richard Morton[2] (2019 – ongoing)
Christopher Short[1] (2021 – ongoing)
Simon Stoneham[2] (2021 - ongoing)
Alice Collins[2] (2019 – ongoing)
Luca Robinson[2] (2021 – ongoing)
Micaela Mossop[2] (2021 – ongoing)

Post-doctoral fellows
Theresa Wodehouse (2003- 2005)
Nicholas Regamey (2005-2008)
Rossa Brugha (2016-2018)
Andrew Turnbull (2019 – 2020)
Matt Coates (2020-2022)

Grants Awarded

Apr – Nov 2022 **Imperial College London Healthcare NHS Trust - BRC funding** **£40,768**

Pseudomonas aeruginosa virulence and persistence mechanisms as novel therapeutic targets for cystic fibrosis**.**

2021 – 2024 **Cystic Fibrosis Foundation** **£654,962**

Oxygen-enhanced MRI as an outcome measure in cystic fibrosis

2020 – 2024 **Cystic Fibrosis Foundation, Cystic Fibrosis Trust and CF Ireland** **£2,543,968**

Real World Clinical Outcomes with Novel Modulator Therapy Combinations in People with CF (RECOVER)

2020 – 2021 **Cystic Fibrosis Trust** **£50,000**

Exploring the utility of quorum sensing inhibitors and biofilm disruptors on growth and virulence behaviours of Pseudomonas aeruginosa obtained from patients with Cystic Fibrosis

2020 – 2024 **NIHR** **£80,000**

NIHR Senior Investigator Award

2020 – 2022 **EPSRC** **£850,000**

The Idealized Lung Clearance Index

2020 – 2022 **Cystic Fibrosis Trust** **£98,594.93**

The idealised LCI (i-LCI): tuning in on the ‘silent years’ of paediatric CF

2018-2022 **Cystic Fibrosis Trust** **£750,000**

Personalised Approach to Pseudomonas aeruginosa (PAPA)

2018 – 2020 **Vertex Pharmaceuticals** **£56,121**

Children's Follow up Orkambi Real world MBW Study (C-FORMS)

2018-2019 **Cystic Fibrosis Trust** **£50,000**

Exploring the utility of novel ‘antimicrobial resistance breakers’ on strains of Pseudomonas aeruginosa obtained from patients with Cystic Fibrosis

2018-2019 **Cystic Fibrosis Trust** **£20,000**

LCI Training Package for the Cystic Fibrosis Clinical Trials Accelerator Platform (CTAP)

2017-2022 Department of Health/ Wellcome Trust £550,000
First-in-human trial of an optimised lentiviral vector for cystic fibrosis gene therapy

2016-2019 Cystic Fibrosis Trust £81,312.58 (3 awards)
Exploring the antibacterial activity of Glatiramer acetate on strains of Pseudomonas aeruginosa obtained from patients with Cystic Fibrosis at varying stages of disease progression.

2017-2020 Cystic Fibrosis Trust £90,000
RAPID point-of-care infection detection and antibiotic-resistance TESTing enabled with laser-patterned microfluidic devices (RAPID-TEST)

2017-2020 EPSRC £96,223.44
RAPID point-of-care infection detection and antibiotic-resistance TESTing enabled with laser-patterned microfluidic devices (RAPID-TEST)

2017 Cystic Fibrosis Trust £4,497

Grant Award for Cystic Fibrosis Clinical Trials Database Information Support

2016-2018 Cystic Fibrosis Trust £372,724.22
CLIMB-CF: Clinical Monitoring and Biomarkers to stratify severity and predict outcomes in children with cystic fibrosis

2016-17 Cystic Fibrosis Trust £28,409
SmartCareCFKids: Home monitoring for the prompt recognition of Pulmonary exacerbations (PEx)in children with cystic fibrosis.

2016-2017 British Lung Foundation £25,000

The role of bacterial biofilms in children with chronic suppurative lung diseases

2016-2021 National Institutes for Health Research £283,606
Improving Outcome Measures For Physiotherapy Trials of Airway Clearance in Adult Cystic Fibrosis

2016 National Institutes for Health Research £69,555
Stratifying disease severity in paediatric cystic fibrosis: identifying high risk children in different age groups - Dr Claire Edmondson Fellowship.

2015-2016 Imperial College Antimicrobial Research Collaborative; Early Career Fellowship £57,832.49

Subverting bacterial c-di-GMP signalling to fight antimicrobial resistance in the clinic.

2014 Cystic Fibrosis Trust £44,507

Cystic Fibrosis Trust: CF Gene Therapy Consortium core funding 1st tranche

2014-2018 Cystic Fibrosis Trust £750,000

Strategic Research Centre for Pseudomonas Research

2012 – 2014 NIHR through EME scheme £3,073,900

A randomised double-blind placebo-controlled Phase 2B clinical trial of repeated application of gene therapy in patients with cystic fibrosis.

£1,243,400
2012 – 2013 DPFS Medical Research Council £34,146 (extended)

Development of a novel, potent, safe, long-lasting lentivirus-based gene therapy for cystic fibrosis.

2010 – 2011 Phage Special Services £65,547

Industrial Collaboration 'The potential for phage therapy to treat bacterial airway infection in cystic fibrosis'

Royal Brompton Hospital Biomedical
2010 – 2011 Research Unit Pump Priming Fund £9,450

Volatile organic compounds in exhaled breath as markers of lower airway bacterial infection

2003 – 2011 Cystic Fibrosis Trust Various see below

UK Gene Therapy Consortium Grants:
"Gene Therapy for Cystic Fibrosis: Towards Clinical Reality"

Internal Investigators:	Eric Alton, Jane Davies, Uta Griesenbach
	Depts of Gene Therapy, Paediatric Respiratory Medicine
External Investigators:	David Porteous, Medical Genetics, Edinburgh University
	Steven Hyde, University of Oxford

2003 £761,329	2007 £4,704,675
2004 £2,786,442	2010 £4,784,692 (cancelled in 2011)
2006 £971,464	2011 £1,600,000

Measures of Esteem

- Invited 2023 Royal College of Physicians Croonian lecturer
- NIHR Senior Investigator Award 2020-
- Fellow of the European Respiratory Society 2018-
- Invited Plenary lecturer:
  - NACF Conference 2019
  - European CF Conference: 2012, 2017, 2019
  - International Conference on Paediatric Pulmonology (CIPP) 2017
  - British Thoracic Soc Winter Meeting 2019
  - Australasian CF Conference 2019
  - Talamo Lecturer Award 2019
- NIHR award for commercial trials leadership 2016
- NIHR NW London CRN Awards for: Outstanding PI & Best research team 2019
- Imperial College London Rector's Gold Medal for Research Supervision 2014
- Chair DSMB for multicentre, NIHR-funded clinical trial (ProteKt, 2016-2018)

Leadership roles (current unless stated)

European CF Society

- Deputy President 2020-
- Task Force: Strategic Planning for faster access to new drugs, Lead
- Diagnostic Working Group, Site Lead
- Standardisation Committee, member
- Clinical Trials Network, Principal Investigator
- Lung Clearance Index Core Facility, Lead
- Conference: Scientific Committee (2004, 2012-15); President 2019

CF Trust

- Strategic Advisory Board (2013- 2022), Vice Chair
- Clinical Trials Accelerator Platform Research & Scientific Oversight Committee, Chair (2017-2022)
- CTAP London Network, Lead

British Thoracic Society

- Science & Research Committee (2014- 2022)
- Training & Education Committee (2014- 2020)

National institutes for Health Research

- Imperial Academic Health Sciences Lead on Respiratory Translational Research Collaboration (2019- ongoing

Royal Brompton, Harefield and NHLI

- Biomedical Research Centre, Co-lead Respiratory Theme
- Deputy Head of Section, NHLI, Imperial College London (2019 -ongoing)
- Clinical Research Oversight Committee (2015- ongoing)
- Respiratory Research Committee (2017- ongoing)
- Research Ethics Committee (2007-2010)

UK CF Gene Therapy Consortium

- Strategy Group member

Other

- Medicines Discovery Catapult/ CFT Syndicate for Antimicrobial Resistance: Deputy Chair
- British Paediatric Respiratory Society: Research Committee Chair (2014- 2022)
- lancet (2015- 2020)
- Medicines for Children Research Network: Respiratory and CF CSG (2010-)
- American Thoracic Society Conference Paediatric Assembly, 2007, 2008, 2009

Journal/ Editorial roles

- Deputy Editor Journal of Cystic Fibrosis (2020-ongoing)
- Lancet Respiratory Medicine, Advisory Board (2019- ongoing)
- Associate Editor Thorax (2015- 2020)
- Editorial Board Pediatric Allergy, Immunology and Pulmonology
- Series Editor: New Biology of the Airways, Paediatric Respiratory Reviews

Clinical Advisory Roles

- LifeArc
- PIPE-CF; Strategic Research Centre, Liverpool University
- Vertex Pharmaceuticals
- AbbVie

- Galapagos NV
- Chiesi Limited
- Proteostasis Therapeutics Inc.
- Eloxx Pharmaceuticals Inc.
- Boehringer-Ingelheim Pharma GmbH & Co. KG
- AlgiPharma AS
- Arcturus Therapeuticts
- Flatley Discovery Lab
- Pulmocide
- Novartis
- Enterprise Therapeutics
- Raptor Phamaceuticals Inc.
- ProQR Therapeutics III B.V.
- Bayer AG
- ImevaX GmbH
- Nivalis Therapeutics, Inc.

Review/ referee activities

Grants:

In recent years I have reviewed grant applications for the following bodies:

- National Institutes for Health Research
- Cystic Fibrosis Trust
- Canadian CF Society
- Cystic Fibrosis in Australia
- German Federal Ministry of Education and Research Funding Initiative on Rare Disease Research Consortia
- Italian Research Foundation
- Gilead Research Scholars Program (2017-ongoing)
- Vertex Venture & Innovations Awards (2017-ongoing)

Journal submissions:

I regularly review scientific manuscripts for the major respiratory journals including Lancet Respir Med, Am J Respir Crit Care Med, ERJ, Annals ATS, Pediatr Pulmonol, Thorax, JCF, Chest

Publications *(Google scholar h-index 69; Feb 2023)*

Original Research

Diversity and prevalence of type VI secretion system effectors in clinical *Pseudomonas aeruginosa* isolates. Robinson LA, Collins ACZ, Murphy RA, **Davies JC**, Allsopp LP. *Front Microbiol*. 2023;13:1042505.

Migration is not the perfect answer: How the cross-talk error correction for multiple breath nitrogen washout (MBWN2) parameters differs on directly collected vs. legacy data. Short C, Abkir M, Pinnell S, Proctor O, Saunders CJ, **Davies JC**. *Pediatr Pulmonol*. 2022 Sep 29.

Synergistic Activity of Repurposed Peptide Drug Glatiramer Acetate with Tobramycin against Cystic Fibrosis Pseudomonas aeruginosa. Murphy RA, Coates M, Thrane S, Sabnis A, Harrison J, Schelenz S, Edwards AM, Vorup-Jensen T, **Davies JC**. *Microbiol Spectr*. 2022 Aug 31;10(4):e0081322.

An invisible threat? Aspergillus positive cultures and co-infecting bacteria in airway samples. Hughes DA, Rosenthal M, Cuthbertson L, Ramadan N, Felton I, Simmonds NJ, Loebinger

MR, Price H, Armstrong-James D, Elborn JS, Cookson WO, Moffatt MF, Davies JC. *J Cyst Fibros*. 2022 Jul 21;S1569-1993(22)00627-0.

Efficacy and Safety of Elexacaftor/Tezacaftor/Ivacaftor in Children 6 Through 11 Years of Age with Cystic Fibrosis Heterozygous for F508del and a Minimal Function Mutation: A Phase 3B, Randomized, Placebo-Controlled Study. Mall MA, Brugha R, Gartner S, Legg J, Moeller A, Mondejar-Lopez P, Prais D, Pressler T, Ratjen F, Reix P, Robinson PD, Selvadurai H, Stehling F, Ahluwalia N, Arteaga-Solis E, Bruinsma BG, Jennings M, Moskowitz SM, Noel S, Tian S, Weinstock TG, Wu P, Wainwright CE, **Davies JC**, VX19-445-116 Study Group. *Am J Respir Crit Care Med*. 2022 Jul 11.

Curvilinearity provides additional information to lung clearance index only in a minority of children with early cystic fibrosis lung disease. Irving S, Bayfield K, **Davies JC**, Bush A. *ERJ Open Res*. 2022 Apr 4;8(2):00582-2021.

A Phase 3, open-label, 96-week trial to study the safety, tolerability, and efficacy of tezacaftor/ivacaftor in children ≥ 6 years of age homozygous for F508del or heterozygous for F508del and a residual function CFTR variant. Sawicki GS, Chilvers M, McNamara J, Naehrlich L, Saunders C, Sermet-Gaudelus I, Wainwright CE, Ahluwalia N, Campbell D, Harris RS, Paz-Diaz H, Shih JL, **Davies JC.** *J Cyst Fibros.* 2022 Feb 18:S1569-1993(22)00033-9.

The feasibility of home monitoring of young people with cystic fibrosis: Results from CLIMB-CF. Edmondson C, Westrupp N, Seddon P, Olden C, Wallis C, Dawson C, Brodlie M, Baxter F, McCormick J, MacFarlane S, Rice D, Macleod A, Brooker R, Connon M, Ghayyda S, Blaikie L, Thursfield R, Brown L, Price A, Fleischer E, Itterman J, Hughes D, Barrett P, Surette M, Donnelly C, Mateos-Corral D, Padley G, Wallenburg J, Brownlee K, Alton EWFW, Bush A, **Davies JC**. *J Cyst Fibros*. 2022 Jan;21(1):70-77.

Impact of cross-sensitivity error correction on representative nitrogen-based multiple breath washout data from clinical trials. Robinson PD, Jensen R, Seeto RA, Stanojevic S, Saunders C, Short C, **Davies JC**, Ratjen F. *J Cyst Fibros.* 2021 Sep 12:S1569-1993(21)01374-6.

Comparison of the airway microbiota in children with chronic suppurative lung disease. Ahmed B, Cox MJ, Cuthbertson L, James P, Gardner L, Cookson W, **Davies J** et al. *BMJ Open Respir Res*. 2021 Dec;8(1):e001106.

Clinical pharmacokinetics and dose recommendations for posaconazole gastroresistant tablets in children with cystic fibrosis. Bentley S, **Davies JC** et al. *J Antimicrob Chemother*. 2021 Nov 12;76(12):3247-3254.

Riociguat for the treatment of Phe508del homozygous adults with cystic fibrosis. Derichs N, Taylor-Cousar JL, **Davies JC** et al.*J Cyst Fibros*. 2021 Nov;20(6):1018-1025.

Guiding the rational design of patient-centred drug trials in Cystic Fibrosis: A Delphi study. Dobra R, Elborn JS, Madge S, Allen L, Boeri M, Kee F, Goundry S, Purcell T, Saunders C, **Davies JC**. *J Cyst Fibros*. 2021 Nov;20(6):986-993.

A Short extension to multiple breath washout provides additional signal of distal airway disease in people with CF: A pilot study. Short C, Semple T, Saunders C, Hughes D, Irving S, Gardener L, Rosenthal M, Robinson PD, **Davies JC**. *J Cyst Fibros*. 2022 Jan;21(1):146-154.

Updated guidance on the management of children with cystic fibrosis transmembrane conductance regulator-related metabolic syndrome/cystic fibrosis screen positive, inconclusive diagnosis (CRMS/CFSPID). Barben J, Castellani C, Munck A, **Davies JC** et al. *J Cyst Fibros*. 2021 Sep;20(5):810-819.

Triple Therapy for Cystic Fibrosis Phe508del-Gating and -Residual Function Genotypes. Barry PJ, Mall MA, Álvarez 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; **VX18-445-104 Study Group.** *N Engl J Med*. 2021 Aug 26;385(9):815-825.

Efficacy and safety of inhaled ENaC inhibitor BI 1265162 in patients with cystic fibrosis: BALANCE-CF 1 - a randomised, Phase II study. Goss CH, Fajac I, Jain R, Seibold W, Gupta A, Hsu MC, Sutharsan S, **Davies JC**, Mall MA. *Eur Respir J*. 2021 Aug 12:2100746.

Long-term safety and efficacy of tezacaftor-ivacaftor in individuals with cystic fibrosis aged 12 years or older who are homozygous or heterozygous for Phe508del CFTR (EXTEND): an open-label extension study. Flume PA, Biner RF, Downey DG, Brown C, Jain M, Fischer R, De Boeck K, Sawicki GS, Chang P, Paz-Diaz H, Rubin JL, Yang Y, Hu X, Pasta DJ, Millar SJ, Campbell D, Wang X, Ahluwalia N, Owen CA, Wainwright CE; **VX14-661-110 study group**. *Lancet Respir Med*. 2021 Jul;9(7):733-746.

Long-term safety and efficacy of lumacaftor-ivacaftor therapy in children aged 6-11 years with cystic fibrosis homozygous for the F508del-CFTR mutation: a phase 3, open-label, extension study. Chilvers MA, **Davies JC** et al. *Lancet Respir Med*. 2021 Jul;9(7):721-732.

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. Zemanick ET, Taylor-Cousar JL, **Davies J** et al. *Am J Respir Crit Care Med*. 2021 Jun 15;203(12):1522-1532.

Targeted exhaled breath analysis for detection of Pseudomonas aeruginosa in cystic fibrosis patients. Kos R, Brinkman P, Neerincx AH, Paff T, Gerritsen MG, Lammers A, Kraneveld AD, Heijerman HGM, Janssens HM, **Davies JC** et al. *J Cyst Fibros*. 2021 May 17:S1569-1993(21)00125-9.

Clinical characteristics of Pseudomonas and Aspergillus co-infected cystic fibrosis patients: A UK registry study. Hughes DA, Archangelidi O, Coates M, Armstrong-James D, Elborn SJ, Carr SB, **Davies JC**. *J Cyst Fibros*. 2021 May 3:S1569-1993(21)00117-X.

Pseudomonas aeruginosa in the Cystic Fibrosis Airway: Does It Deserve Its Reputation as a Predatory "Bully"? Hughes DA, Price H, Rosenthal M, **Davies JC**. *Am J Respir Crit Care Med*. 2021 Apr 15;203(8):1027-1030.

Transepithelial nasal potential difference in patients with, and at risk of acute respiratory distress syndrome. Mac Sweeney R, Reddy K, **Davies JC**, Parker M, Kelly B, Elborn JS, Conlon J, Verghis RM, Calfee CS, Matthay MA, Alton EWFW, McAuley DF. *Thorax*. 2021 Apr 22:thoraxjnl-2020-215587.

Colistin kills bacteria by targeting lipopolysaccharide in the cytoplasmic membrane. Sabnis A, Hagart KLH, Klöckner A, Becce M, Evans LE, Furniss RCD, Mavridou DAI, Murphy R, Stevens MM, **Davies JC**, Larrouy-Maumus GJ, Clarke TB, Edwards AM. *Elife*. 2021 Apr 6;10:e65836.

Variability in Bacteriophage and Antibiotic Sensitivity in Serial Pseudomonas aeruginosa Isolates from Cystic Fibrosis Airway Cultures over 12 Months. Martin I, Kenna DTD, Morales S, Alton EWFW, **Davies JC**. *Microorganisms*. 2021 Mar 22;9(3):660.

A Phase 3 Open-Label Study of ELX/TEZ/IVA in Children 6 Through 11 Years of Age With CF and at Least One F508del Allele. 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; VX18-445-106 Study Group. *Am J Respir Crit Care Med*. 2021 Mar 18.

Discrete choice experiment (DCE) to quantify the influence of trial features on the decision to participate in cystic fibrosis (CF) clinical trials. Dobra RA, Boeri M, Elborn S, Kee F, Madge S, **Davies JC**. *BMJ Open*. 2021 Mar 2;11(3):e045803.

Pseudomonas aeruginosa induces p38MAP kinase-dependent IL-6 and CXCL8 release from bronchial epithelial cells via a Syk kinase pathway. Coates MS, Alton EWFW, Rapeport GW, **Davies JC**, Ito K. *PLoS One*. 2021 Feb 1;16(2):e0246050.

Long-term safety and efficacy of lumacaftor-ivacaftor therapy in children aged 6-11 years with cystic fibrosis homozygous for the F508del-CFTR mutation: a phase 3, open-label, extension study. Chilvers MA, **Davies JC**, Milla , Tian S, Han Z, Cornell AG, Owen CA, Ratjen F. *Lancet Respir Med*. 2021 Jan 28;S2213-2600(20)30517-8.

A phase 3, double-blind, parallel-group study to evaluate the efficacy and safety of tezacaftor in combination with ivacaftor in participants 6 through 11 years of age with cystic fibrosis homozygous for F508del or heterozygous for the F508del-CFTR mutation and a residual function mutation. **Davies JC**, Sermet-Gaudelus I, Naehrlich L, Harris RS, Campbell D, Ahluwalia N, Short C, Haseltine E, Panorchan P, Saunders C, Owen CA, Wainwright CE; VX16-661-115 Investigator Group. *J Cyst Fibros*. 2021 Jan;20(1):68-77.

Pseudomonas aeruginosa in the CF Airway: Does it Deserve its Reputation as a Predatory 'Bully'? Hughes DA, Price H, Rosenthal M**, Davies JC**. *Am J Respir Crit Care Med*. 2020 Dec 2.

Multiple breath washout in bronchiectasis clinical trials: is it feasible? O'Neill K, Ferguson K, Cosgrove D, Tunney MM, De Soyza A, Carroll M, Chalmers JD, Gatheral T, Hill AT, Hurst JR, Johnson C, Loebinger MR, Angyalosi G, Haworth CS, Jensen R, Ratjen F, Saunders C, Short C, **Davies JC**, Elborn JS, Bradley JM. *ERJ Open Res*. 2020 Oct 13;6(4):00363-2019.

Ivacaftor in Infants Aged 4 to <12 Months With Cystic Fibrosis and a Gating Mutation: Results of a 2-Part Phase 3 Clinical Trial. **Davies JC**, Wainwright CE, Sawicki GS et al; ARRIVAL Study Group. *Am J Respir Crit Care Med*. 2020 Oct 7.

Response of Pseudomonas aeruginosa to the innate immune system-derived oxidants hypochlorous acid and hypothiocyanous acid. Farrant KV, Spiga L, **Davies JC**, Williams HD. *J Bacteriol*. 2020 Oct 26:JB.00300-20.

Ivacaftor in People With Cystic Fibrosis and a 3849+10kb C →T or D1152H Residual Function Mutation. Kerem E, Cohen-Cymberknoh M, Tsabari R, Wilschanski M, Reiter J, Shoseyov D, Gileles-Hillel A, Pugatsch T, **Davies JC** et al. *Ann Am Thorac Soc*. 2020 Oct 23.

Inhaled dry powder alginate oligosaccharide in cystic fibrosis: a randomised, double-blind, placebo-controlled, crossover phase 2b study. van Koningsbruggen-Rietschel S, **Davies JC** et al. *ERJ Open Res*. 2020 Oct 19;6(4):00132-2020.

Longitudinal immune profiling reveals key myeloid signatures associated with COVID-19. Mann ER, Menon M, Knight SB, Konkel JE, Jagger C, Shaw TN, Krishnan S, Rattray M, Ustianowski A, Bakerly ND, Dark P, Lord G, Simpson A, Felton T, Ho LP; **NIHR Respiratory TRC**, Feldmann M; CIRCO, Grainger JR, Hussell T. *Sci Immunol*. 2020 Sep 17;5(51):eabd6197.

A phase 3, double-blind, parallel-group study to evaluate the efficacy and safety of tezacaftor in combination with ivacaftor in participants 6 through 11 years of age with cystic fibrosis homozygous for F508del or heterozygous for the F508del-CFTR mutation and a residual function mutation. **Davies JC**, Sermet-Gaudelus I, Naehrlich L et al; VX16-661-115 Investigator Group. *J Cyst Fibros*. 2020 Sep 21:S1569-1993(20)30811-0.

Combination antifungal therapy for Scedosporium species in cystic fibrosis. Bentley S, **Davies JC**, Carr SB, Balfour-Lynn IM. *Pediatr Pulmonol*. 2020 Aug;55(8):1993-1995.

Evaluation of a multiple breath nitrogen washout system in children.
Isaac SM, Jensen R, Anagnostopoulou P, **Davies JC** et al. *Pediatr Pulmonol*. 2020 Aug;55(8):2108-2114.

Inflammation in children with neuromuscular disorders and sleep disordered breathing. Trucco F, Carruthers E, Davies JC, Simonds AK, Bush A, Tan HL. *Sleep Med*. 2020 Aug;72:118-121.

Whole-Gene Sequencing of CFTR Reveals a High Prevalence of the Intronic Variant c.3874-4522A>G in Cystic Fibrosis. Morris-Rosendahl DJ, Edwards M, McDonnell MJ, John S, Alton EWFW, **Davies JC**, Simmonds NJ. *Am J Respir Crit Care Med*. 2020 Jun 1;201(11):1438-1441.

Simultaneous sulfur hexafluoride and nitrogen multiple-breath washout (MBW) to examine inherent differences in MBW outcomes. Bayfield, Horsley A, Alton E, Irving S, Bush A, **Davies JC**. *ERJ Open Res.* 2019 Nov 4;5(4).

Insights into the variability of nasal potential difference, a biomarker of CFTR activity. Kyrilli S, Henry T, Wilschanski M, Fajac I, **Davies JC**, Jais JP, Sermet-Gaudelus I. *J Cyst Fibros*. 2019 Nov 4. pii: S1569-1993(19)30899-9.

Abnormal pro-gly-pro pathway and airway neutrophilia in pediatric cystic fibrosis. Turnbull AR, Pyle CJ, Patel DF, Jackson PL, Hilliard TN, Regamey N, Tan H-L, Brown S, Thursfield R, Short C, Mc Fie M, Alton EWFW, Gaggar A, Blalock JE, Lloyd CM, Bush A, **Davies JC**, Snelgrove RJ. *J Cyst Fibros*. 2020 Jan;19(1):40-48.

Cephalosporin nitric oxide-donor prodrug DEA-C3D disperses biofilms formed by clinical cystic fibrosis isolates of Pseudomonas aeruginosa. Soren O, Rineh A, Silva D, Cai Y, Howlin R, Allan R, Feelisch M, **Davies JC**, Connett G, Faust S, Kelso M, Webb J. *J Antimicrob Chemother* 2020 Jan 1;75(1):117-125.

Integrating the multiple breath washout test into international multicentre trials. Saunders C, Jensen R, Robinson PD, Stanojevic S, Klingel M, Short C, **Davies JC**, Ratjen F. *J Cyst Fibros*. 2019 Nov 23. pii: S1569-1993(19)30968-3.

Training dogs to differentiate *Pseudomonas aeruginosa* from other cystic fibrosis bacterial pathogens: not to be sniffed at? **Davies JC**, Alton E, Simbo A, Murphy R, Seth I, Williams K, Somerville M, Jolly L, Morant S, Guest C. *Eur Respir J*. 2019 Nov 7;54(5).

GLPG1837, a CFTR potentiator, in p.Gly551Asp (G551D)-CF patients: An open-label, single-arm, phase 2a study (SAPHIRA1). **Davies JC**, Van de Steen O, van Koningsbruggen-Rietschel S, Drevinek P, Derichs N, McKone EF, Kanters D, Allamassey L, Namour F, de Kock H, Conrath K. *J Cyst Fibros.* 2019 Sep;18(5):693-699 .

An open-label extension study of ivacaftor in children with CF and a CFTR gating mutation initiating treatment at age 2-5 years (KLIMB). Rosenfeld M, Cunningham S, Harris WT, Lapey A, Regelmann WE, Sawicki GS, Southern KW, Chilvers M, Higgins M, Tian S, Cooke J, **Davies JC**, KLIMB study group. *J Cyst Fibros* 2019. 2019 Nov;18(6):838-843.

Longitudinal development of the airway microbiota in infants with cystic fibrosis. Ahmed B, Cox MJ, Cuthbertson L, James P, Cookson WOC, **Davies JC** et al. *Sci Rep*. 2019 Mar 26;9(1):5143.

VX-659-Tezacaftor-Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles. **Davies JC** et al. *N Engl J Med*. 2018 Oct 25;379(17):1599-1611.

Comparison of the upper and lower airway microbiota in children with chronic lung diseases. Ahmed B, Cox MJ, Cuthbertson L, James PL, Cookson WOC, **Davies JC** et al. *PLoS One*. 2018 Aug 2;13(8):e0201156.

Metabolic Phenotyping and Strain Characterisation of Pseudomonas aeruginosa Isolates from Cystic Fibrosis Patients Using Rapid Evaporative Ionisation Mass Spectrometry. Bardin EE, Cameron SJS, Perdones-Montero A, Hardiman K, Bolt F, Alton EWFW, Bush A, **Davies JC**, Takáts Z. *Sci Rep*. 2018 Jul 19;8(1):10952.

Ivacaftor treatment of cystic fibrosis in children aged 12 to <24 months and with a CFTR gating mutation (ARRIVAL): a phase 3 single-arm study. Rosenfeld M, Wainwright CE, Higgins M, Wang LT, McKee C, Campbell D, Tian S, Schneider J, Cunningham S, **Davies JC**; ARRIVAL study group. *Lancet Respir Med*. 2018 Jul;6(7):545-553.

Impact of T2R38 Receptor Polymorphisms on Pseudomonas aeruginosa Infection in Cystic Fibrosis. Turnbull AR, Murphy R, Behrends V, Lund-Palau H, Simbo A, Mariveles M, Alton EW, Bush A, Shoemark A, **Davies JC**. *Am J Respir Crit Care Med*. 2018 Jun 15;197(12):1635-1638.

Pooling of bronchoalveolar lavage in children with cystic fibrosis does not adversely affect the microbiological yield or sensitivity in detecting pulmonary inflammation. McNally P, O'Rourke J, Fantino E, Chacko A, Pabary R, Turnbull A, Grant T, O'Sullivan N, Wainwright C, Linnane B, **Davies JC**, Sly PD. *J Cyst Fibros*. 2018 May;17(3):391-399.

Children with cystic fibrosis demonstrate no respiratory immunological, infective or physiological, consequences of vitamin D deficiency. Thursfield RM, Naderi K, Leaver N, Rosenthal M, Alton EWFW, Bush A, **Davies JC**. *J Cyst Fibros*. 2018 Apr 6. pii: S1569-1993(18)30070-5.

Tezacaftor/Ivacaftor in Subjects with Cystic Fibrosis and F508del/F508del-CFTR or F508del/G551D-CFTR. Donaldson SH, Pilewski JM, Griese M, Cooke J, Viswanathan L, Tullis E, **Davies JC**, Lekstrom-Himes JA, Wang LT. *Am J Respir Crit Care Med*. 2018 Jan 15;197(2):214-224.

Recovery of lung function following a pulmonary exacerbation in patients with cystic fibrosis and the G551D-CFTR mutation treated with ivacaftor. Flume PA, Wainwright CE, Elizabeth

Tullis D, Rodriguez S, Niknian M, Higgins M, **Davies JC**, Wagener JS. *J Cyst Fibros*. 2018 Jan;17(1):83-88.

A Cell-Free Biosensor for Detecting Quorum Sensing Molecules in P. aeruginosa-Infected Respiratory Samples. Wen KY, Cameron L, Chappell J, Jensen K, Bell DJ, Kelwick R, Kopniczky M, **Davies JC**, Filloux A, Freemont PS. *ACS Synth Biol*. 2017 Dec 15;6(12):2293-2301.

Tezacaftor-Ivacaftor in Residual-Function Heterozygotes with Cystic Fibrosis. Rowe SM, Daines C, Ringshausen FC, Kerem E, Wilson J, Tullis E, Nair N, Simard C, Han L, Ingenito EP, Mckee, C, Lekstrom-Himes J, **Davies JC**. N Engl *J Med*. 2017 Nov 23;377(21):2024-2035.

The Immunomodulatory Drug Glatiramer Acetate is Also an Effective Antimicrobial Agent that Kills Gram-negative Bacteria. Hill Christiansen S, Murphy RA, Juul-Madsen K, Fredborg M, Lykke Hvam M, Axelgaard E, Skovdal SM, Meyer RL, Skov Sørensen UB, Möller A, Nyengaard JR, Nørskov-Lauritsen N, Wang M, Gadjeva M, Howard KA, **Jane C. Davies**, Petersen E, Vorup-Jensen T. *Sci Rep*. 2017 Nov 15;7(1):15653.

Efficacy and safety of lumacaftor and ivacaftor in patients aged 6-11 years with cystic fibrosis homozygous for F508del-CFTR: a randomised, placebo-controlled phase 3 trial. Ratjen F, Hug C, Marigowda G, Tian S, Huang X, Stanojevic S, Milla CE, Robinson PD, Waltz D, Davies JC; VX14-809-109 investigator group. Lancet Respir Med. 2017 Jul;5(7):557-567.

Preparation for a first-in-man lentivirus trial in patients with cystic fibrosis. Eric W F W Alton, Jeffery M Beekman, A Christopher Boyd, June Brand, Marianne S Carlon, Mary M Connolly, Mario Chan, Sinead Conlon, Heather E Davidson, **Jane C Davies** et al. *Thorax*. 2017 Feb; 72(2): 137–147.

Diagnosis of Cystic Fibrosis in Screened Populations. Farrell PM, White TB, Howenstine MS, Munck A, Parad RB, Rosenfeld M, Sommerburg O, Accurso FJ, **Davies JC**, Rock MJ, Sanders DB, Wilschanski M, Sermet-Gaudelus I, Blau H, Gartner S, McColley SA. *J Pediatr*. 2017 Feb;181S:S33-S44.e2.

Variability of sweat chloride concentration in subjects with cystic fibrosis and G551D mutations. Vermeulen F, Le Camus C, **Davies JC**, Bilton D, Milenković D, De Boeck K. *J Cyst Fibros*. 2017 Jan;16(1):36-40.

Does mass spectrometric breath analysis detect Pseudomonas aeruginosa in cystic fibrosis? Pabary R, Huang J, Kumar S, Alton EW, Bush A, Hanna GB, **Davies JC**. *Eur Respir J*. 2016 Mar;47(3):994-7.

Safety, pharmacokinetics, and pharmacodynamics of ivacaftor in patients aged 2-5 years with cystic fibrosis and a CFTR gating mutation (KIWI): an open-label, single-arm study. **Davies JC**, Cunningham S, Harris WT, Lapey A, Regelmann WE, Sawicki GS, Southern KW, Robertson S, Green Y, Cooke J, Rosenfeld M; KIWI Study Group. *Lancet Respir Med*. 2016 Feb;4(2):107-15.

Repeated nebulisation of non-viral CFTR gene therapy in patients with cystic fibrosis: a randomised, double-blind, placebo-controlled, phase 2b trial. Alton EW, Armstrong DK, Ashby D, Bayfield KJ, Bilton D, Bloomfield EV, Boyd AC, Brand J, Buchan R, Calcedo R, Carvelli P, Chan M, Cheng SH, Collie DD, Cunningham S, Davidson HE, Davies G, **Davies JC** et al. *Lancet Respir Med*. 2015 Sep;3(9):684-91.
And in *Southampton (UK): NIHR Journals Library*; 2016 Jul.

A Phase I/IIa Safety and Efficacy Study of Nebulized Liposome-mediated Gene Therapy for Cystic Fibrosis Supports a Multidose Trial. Alton EW, Boyd AC, Porteous DJ, Davies G, **Davies JC**, Griesenbach U, Higgins TE, Gill DR, Hyde SC, Innes JA; UK Cystic Fibrosis Gene Therapy Consortium *. *Am J Respir Crit Care Med*. 2015 Dec 1;192(11):1389-92.

The reproducibility and responsiveness of the lung clearance index in bronchiectasis. Grillo L, Irving S, Hansell DM, Nair A, Annan B, Ward S, Bilton D, Main E, **Davies J**, Bush A, Wilson R, Loebinger MR. *Eur Respir J*. 2015 Dec;46(6):1645-53.

Multiple breath washouts in children can be shortened without compromising quality. Ahmad F, Irving S, Alton E, **Davies JC**, Macleod K, Rosenthal M, Saunders C,Bush A, Saglani S, Fleming L. *Eur Respir J*. 2015 Dec;46(6):1814-6.

Antipseudomonal Bacteriophage Reduces Infective Burden and Inflammatory Response in Murine Lung. Pabary R, Singh C, Morales S, Bush A, Alshafi K, Bilton D, Alton EW, Smithyman A, **Davies JC**. *Antimicrob Agents Chemother*. 2015 Nov 16;60(2):744-51.

Lumacaftor-Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508del CFTR. Wainwright CE, Elborn JS, Ramsey BW, Marigowda G, Huang X, Cipolli M, Colombo C, **Davies JC**, De Boeck K, Flume PA, Konstan MW, McColley SA, McCoy K, McKone EF, Munck A, Ratjen F, Rowe SM, Waltz D, Boyle MP; TRAFFIC and TRANSPORT StudyGroups. *N Engl J Med*. 2015 Oct 29;373(18):1783.

Lung clearance index in cystic fibrosis subjects treated for pulmonary exacerbations. Sonneveld N, Stanojevic S, Amin R, Aurora P, **Davies J**, Elborn JS, Horsley A, Latzin P, O'Neill K, Robinson P, Scrase E, Selvadurai H, Subbarao P, Welsh L, Yammine S, Ratjen F. *Eur Respir J*. 2015 Oct;46(4):1055-64.

Increased nuclear suppressor of cytokine signaling 1 in asthmatic bronchial epithelium suppresses rhinovirus induction of innate interferons. Gielen V, Sykes A, Zhu J, Chan B, Macintyre J, Regamey N, Kieninger E, Gupta A, Shoemark A, Bossley C, **Davies J,** Saglani S, Walker P, Nicholson SE, Dalpke AH, Kon OM, Bush A, Johnston SL, Edwards MR. *J Allergy Clin Immunol*. 2015 Jul;136(1):177-188.

Long-term safety and efficacy of ivacaftor in patients with cystic fibrosis who have the Gly551Asp-CFTR mutation: a phase 3, open-label extension study (PERSIST). McKone EF, Borowitz D, Drevinek P, Griese M, Konstan MW, Wainwright C, Ratjen F, Sermet-Gaudelus I, Plant B, Munck A, Jiang Y, Gilmartin G, **Davies JC**;VX08-770-105 (PERSIST) Study Group. *Lancet Respir Med*. 2014 Nov;2(11):902-910.

Cyanide levels found in infected cystic fibrosis sputum inhibit airway ciliary function. Nair C, Shoemark A, Chan M, Ollosson S, Dixon M, Hogg C, Alton EW, **Davies JC**, Williams HD. *Eur Respir J*. 2014 Nov;44(5):1253-61.

Ataluren for the treatment of nonsense-mutation cystic fibrosis: a randomised, double-blind, placebo-controlled phase 3 trial. Kerem E, Konstan MW, De Boeck K, Accurso FJ, Sermet-Gaudelus I, Wilschanski M, Elborn JS, Melotti P, Bronsveld I, Fajac I, Malfroot A, Rosenbluth DB, Walker PA, McColley SA, Knoop C, Quattrucci S, Rietschel E, Zeitlin PL, Barth J, Elfring GL, Welch EM, Branstrom A, Spiegel RJ, Peltz SW, Ajayi T, Rowe SM; **Cystic Fibrosis Ataluren Study Group**. *Lancet Respir Med*. 2014 Jul;2(7):539-47.

Toxicology study assessing efficacy and safety of repeated administration of lipid/DNA complexes to mouse lung. Alton EW, Boyd AC, Cheng SH, **Davies JC,** Davies LA, Dayan A,

Gill DR, Griesenbach U, Higgins T, Hyde SC, Innes JA, McLachlan G, Porteous D, Pringle I, Scheule RK, Sumner-Jones S. *Gene Ther.* 2014 Jan;21(1):89-95.

Is chest CT useful in newborn screened infants with cystic fibrosis at 1 year of age? Thia LP, Calder A, Stocks J, Bush A, Owens CM, Wallis C, Young C, Sullivan Y, Wade A, McEwan A, Brody AS; **London Cystic Fibrosis Collaboration**. *Thorax*. 2014 Apr;69(4):320-7.

Nasal potential difference measurements in diagnosis of cystic fibrosis: An international survey. Naehrlich L, Ballmann M, **Davies J**, Derichs N, Gonska T, Hjelte L, van Konigsbruggen-Rietschel S, Leal T, Melotti P, Middleton P, Tümmler B, Vermeulen F, Wilschanski M; on behalf of the ECFS Diagnostic Network Working Group. *J Cyst Fibros*. 2014 Jan;13(1):24-8.

The safety profile of a cationic lipid-mediated cystic fibrosis gene transfer agent following repeated monthly aerosol administration to sheep. Alton EW, Baker A, Baker E, Boyd AC, Cheng SH, Coles RL, Collie DD, Davidson H, **Davies JC**, Gill DR, Gordon C, Griesenbach U, Higgins T, Hyde SC, Innes JA, McCormick D, McGovern M, McLachlan G, Porteous DJ, Pringle I, Scheule RK, Shaw DJ, Smith S, Sumner-Jones SG, Tennant P, Vrettou C. *Biomaterials*. 2013 Dec;34(38):10267-77.

Assessment of clinical response to ivacaftor with lung clearance index in cystic fibrosis patients with a G551D-CFTR mutation and preserved spirometry: a randomised controlled trial. **Davies J**, Sheridan H, Bell N, Cunningham S, Davis SD, Elborn JS, Milla CE, Starner TD, Weiner DJ, Lee P-S, Ratjen R. *Lancet Respir Med.* 2013 Oct;1(8):630-8.
Lung clearance index and high-resolution computed tomography scores in primary ciliary dyskinesia. Irving SJ, Ives A, Davies G, Donovan J, Edey AJ, Gill SS, Nair A, Saunders C, Wijesekera NT, Alton EW, Hansell D, Hogg C, **Davies JC**, Bush A. *Am J Respir Crit Care Med.* 2013 Sep 1;188(5):545-9.

Self-Reactive CFTR T Cells in Humans: Implications for Gene Therapy. Calcedo R, Griesenbach U, Dorgan DJ, Soussi S, Boyd AC, **Davies JC**, Higgins TE, Hyde SC, Gill DR, Innes JA, Porteous DJ, Alton EW, Wilson JM, Limberis MP. *Hum Gene Ther Clin Dev.* 2013 Sep;24(3):108-15.

Impaired innate interferon responses to rhinovirus in severe atopic asthmatic children. Edwards MR, Regamey N, Varielle M, Kieninger E, Gupta A, Shoemark A, Saglani S, Sykes A, Macintyre J, Davies J, Bossley C, Bush A, Johnston SL. *Mucosal Immunol*. 2013 Jul;6(4):797-806.

Efficacy and Safety of Ivacaftor in Patients Aged 6 to 11 Years with Cystic Fibrosis with a G551D Mutation. **Davies JC**, Wainwright CE, Canny GJ, Chilvers MA, Howenstine MS, Munck A, Mainz JG, Rodriguez S, Li H, Yen K, Ordoñez C, Ahrens R; on behalf of the VX08-770-103 (ENVISION) Study Group. *Am J Respir Crit Care Med. 2013* Jun 1;187(11):1219-25.

Changes in physiological, functional and structural markers of cystic fibrosis lung disease with treatment of a pulmonary exacerbation. Horsley AR*, **Davies JC***, Gray RD, Macleod KA, Donovan J, Aziz ZA, Bell NJ, Rainer M, Mt-Isa S, Voase N, Dewar MH, Saunders C, Gibson JS, Parra-Leiton J, Larsen MD, Jeswiet S, Soussi S, Bakar Y, Meister MG, Tyler P, Doherty A, Hansell DM, Ashby D, Hyde SC, Gill DR, Greening AP, Porteous DJ, Innes JA, Boyd AC, Griesenbach U, Cunningham S, Alton EW. *Thorax.* 2013 Jun;68(6):532-9. Joint first authors*.

A molecular comparison of microbial communities in bronchiectasis and cystic fibrosis. Duff RM, Simmonds NJ, **Davies JC**, Wilson R, Alton EW, Pantelidis P, Cox MJ, Cookson WOCM, Bilton D, Moffatt MF. *Eur Respir J.* 2013 Apr;41(4):991-3.

High rhinovirus burden in lower airways of children with cystic fibrosis. Kieninger E, Singer F, Tapparel C, Alves MP, Latzin P, Tan HL, Bossley C, Casaulta C, Bush A, **Davies JC**, Kaiser L, Regamey N. *Chest.* 2013 Mar; 143(3):782-90.

Assessment of F/HN-Pseudotyped Lentivirus as a Clinically Relevant Vector for Lung Gene Therapy. Griesenbach U, Inoue M, Meng C, Farley R, Chan M, Newman NK, Brum A, You J, Kerton A, Shoemark A, Boyd AC, **Davies JC**, Higgins TE, Gill DR, Hyde SC, Innes JA, Porteous DJ, Hasegawa M, Alton EW. *Am J Respir Crit Care Med.* 2012 Nov 1;186(9):846-56.

Novel keto-phospholipids are generated by monocytes and macrophages, detected in Cystic Fibrosis, and activate peroxisome proliferator-activated receptor-γ. Hammond VJ, Morgan AH, Lauder SN, Thomas CP, Brown S, Freeman BA, Lloyd C, **Davies J**, Bush A, Levonen AL, Kansanen E, Villacorta L, Chen YE, Porter N, Garcia Diaz YM, Schopfer F, O'Donnell VB. *J Biol Chem*. 2012 Dec 7;287(50):41651-66.

Distinct patterns of inflammation in the airway lumen and bronchial mucosa of children with cystic fibrosis. Regamey N, Tsartsali L, Hilliard TN, Fuchs O, Tan H, Zhu J, Qiu Y-S, Alton EWFW, Jeffery PK, Bush A, **Davies JC.** *Thorax* 2012 Feb;67:164-70.

A CFTR potentiator in patients with cystic fibrosis and the G551D mutation. Ramsey BW, **Davies J**, McElvaney NG, Tullis E, Bell SC, Dřevínek P, Griese M, McKone EF, Wainwright CE, Konstan MW, Moss R, Ratjen F, Sermet-Gaudelus I, Rowe SM, Dong Q, Rodriguez S, Yen K, Ordoñez C, Elborn JS; VX08-770-102 Study Group. *N Engl J Med*. 2011 Nov 3;365(18):1663-72.

Differential global gene expression in cystic fibrosis nasal and bronchial epithelium. Ogilvie V, Passmore M, Hyndman L, Jones L, Stevenson B, Wilson A, Davidson H, Kitchen RR, Gray RD, Shah P, Alton EW, **Davies JC**, Porteous DJ, Boyd AC. *Genomics*. 2011 Nov;98(5):327-36.

Pre-clinical evaluation of three non-viral gene transfer agents for cystic fibrosis after aerosol delivery to the ovine lung. McLachlan G, Davidson H, Holder E, Davies LA, Pringle IA, Sumner-Jones SG, Baker A, Tennant P, Gordon C, Vrettou C, Blundell R, Hyndman L, Stevenson B, Wilson A, Doherty A, Shaw DJ, Coles RL, Painter H, Cheng SH, Scheule RK, **Davies JC**, Innes JA, Hyde SC, Griesenbach U, Alton EW, Boyd AC, Porteous DJ, Gill DR, Collie DD. *Gene Ther.* 2011 Oct;18(10):996-1005.

The th17 pathway in cystic fibrosis lung disease. Tan HL, Regamey N, Brown S, Bush A, Lloyd CM, **Davies JC**. *Am J Respir Crit Care Med.* 2011Jul 15;184(2):252-8.

Bronchoscopy in Cystic Fibrosis Infants Diagnosed by Newborn Screening. Stafler P, **Davies JC**, Balfour-Lynn IM, Rosenthal M, Bush A. *Pediatr Pulmonol*. 2011 Jul;46(7):696-700.

Cystic fibrosis and survival to 40 years: a study of CFTR function. Simmonds NJ, D'Souza L, Roughton M, Alton EW, **Davies JC**, Hodson ME. *Eur Respir J*. 2011 May;37(5):1076-82.

Quantification of Periciliary Fluid (PCL) Height in Human Airway Biopsies is Feasible, but not Suitable as a Biomarker. Griesenbach U, Soussi S, Larsen MB, Casamayor I, Dewar A, Regamey N, Bush A, Shah PL, **Davies JC,** Alton EW. *Am J Respir Cell Mol Biol*. 2011 Mar;44(3):309-15.

Lung clearance index at 4 years predicts subsequent lung function in children with cystic fibrosis. Aurora P, Stanojevic S, Wade A, Oliver C, Kozlowska W, Lum S, Bush A, Price J, Carr SB, Shankar A, Stocks J; **London Cystic Fibrosis Collaboration**. *Am J Respir Crit Care Med.* 2011 Mar 15;183(6):752-8.

Limitations of the Murine Nose in the Development of Non-viral Airway Gene Transfer.Griesenbach U, Sumner-Jones SG, Holder E, Munkonge FM, Wodehouse T, Smith SN, Wasowicz MY, Pringle I, Casamayor I, Chan M, Coles R, Cornish N, Dewar A, Doherty A, Farley R, Green AM, Jones BL, Larsen MD, Lawton AE, Manvell M, Painter H, Singh C, Somerton L, Stevenson B, Varathalingam A, Siegel C, Scheule RK, Cheng SH, **Davies JC**, Porteous DJ, Gill DR, Boyd AC, Hyde SC, Alton EW. *Am J Respir Cell Mol Biol.* 2010 Jul;43(1):46-54.

Disordered microbial communities in asthmatic airways. Hilty M, Burke C, Pedro H, Cardenas P, Bush A, Bossley C, **Davies J,** Ervine A, Poulter L, Pachter L, Moffatt MF, Cookson WO. *PLoS One*. 2010 Jan 5;5(1):e8578.

An immunocytochemical assay to detect human CFTR expression following gene transfer. Davidson H, Wilson A, Gray RD, Horsley A, Pringle IA, McLachlan G, Nairn AC, Stearns C, Gibson J, Holder E, Jones L, Doherty A, Coles R, Sumner-Jones SG, Wasowicz M, Manvell M, Griesenbach U, Hyde SC, Gill DR, **Davies J**, Collie DD, Alton EW, Porteous DJ, Boyd AC. *Mol Cell Probes*. 2009 Dec;23(6):272-80.

Mannose-binding lectin is present in the infected airway: a possible pulmonary defence mechanism. Fidler KJ, Hilliard TN, Bush A, Johnson M, Alton E, Geddes D, Klein N, Turner M, **Davies J.** *Thorax*. 2009 Feb;64(2):150-5.

Time required to obtain endobronchial biopsies in children during fiberoptic bronchoscopy. Regamey N, Balfour-Lynn I, Rosenthal M, Hogg C, Bush A, **Davies JC.** *Pediatr Pulmonol.* 2009 Jan;44(1):76-9.

Pseudomonas aeruginosa, cyanide accumulation and lung function in CF and non-CF bronchiectasis patients. Ryall B, **Davies JC,** Wilson R, Shoemark A, Williams HD. *Eur Respir J.* 2008 Sep;32(3):740-7.

Biomarkers for cystic fibrosis lung disease: Application of SELDI-TOF mass spectrometry to BAL fluid. Macgregor G, Gray RD, Hilliard TN, Imrie M, Boyd AC, Alton EW, Bush A, **Davies JC,** Innes JA, Porteous DJ, Greening AP. *J Cyst Fibros.* 2008 Sept;7(5):352-8.

CpG-free plasmids confer reduced inflammation and sustained pulmonary gene expression. Hyde SC, Pringle IA, Abdullah S, Lawton AE, Davies LA, Varathalingam A, Nunez-Alonso G, Green AM, Bazzani RP, Sumner-Jones SG, Chan M, Li H, Yew NS, Cheng SH, Christopher Boyd A, **Davies JC**, Griesenbach U, Porteous DJ, Sheppard DN, Munkonge FM, Alton EW, Gill DR. *Nat Biotechnol.* 2008 May;26(5):549-51.

Nasal Abnormalities in Cystic Fibrosis Mice Independent of Infection and Inflammation. Hilliard T, Zhu J, Farley R, Escudero-Garcia S, Jeffery PK, Griesenbach U, Bush A**, Davies JC**, Alton EWFW. *Am J Respir Cell Molec Biol.* 2008 Jul; 39(1):19-25.

Lung function from infancy to the preschool years after clinical diagnosis of cystic fibrosis. Kozlowska WJ, Bush A, Wade A, Aurora P, Carr SB, Castle RA, Hoo AF, Lum S, Price J,

Ranganathan S, Saunders C, Stanojevic S, Stroobant J, Wallis C, Stocks J; **London Cystic Fibrosis Collaboration**. *Am J Respir Crit Care Med*. 2008 Jul 1;178(1):42-9.

Increased airway smooth muscle mass in children with asthma, cystic fibrosis and bronchiectasis. Regamey N, Ochs M, Hilliard TN, Mühlfeld C, Cornish N, Fleming L, Saglani S, Alton EWFW, Bush A, Jeffery PK, **Davies JC**. *Thorax*. 2008 Apr 15; 177(8):837-43.

Lung clearance index is a sensitive, repeatable and practical measure of airways disease in adults with cystic fibrosis. Horsley AR, Gustafsson PM, Macleod K, Saunders CJ, Greening AP, Porteous D, **Davies J,** Cunningham S, Alton E, Innes JA. *Thorax*. 2008 Feb;63(2):135-40.

Endobronchial biopsy in childhood.Regamey N, Hilliard TN, Saglani S, Zhu J, Balfour-Lynn IM, Rosenthal M, Jeffery PK, Alton EW, Bush A, **Davies JC**. *Chest*. 2008 Jan;133(1):312; author reply 313.

Airway remodelling in children with cystic fibrosis. Hilliard TN, Regamey N, Shute J, Nicholson A, Alton EW, Bush A, **Davies JC.** *Thorax*. 2007 Dec;62(12):1074-80.

Bronchoscopy following diagnosis with cystic fibrosis. Hilliard TN, Sukhani S, Francis J, Madden N, Rosenthal M, Balfour-Lynn I, Bush A, **Davies J.** *Arch Dis Child.* 2007 Oct;92(10):898-9.

Quality, size and composition of paediatric endobronchial biopsies in cystic fibrosis. Regamey N, Hilliard TN, Saglani S, Zhu J, Scallan M, Balfour-Lynn IM,Rosenthal M, Jeffery PK, Alton EW, Bush A, **Davies JC.** *Chest*. 2007 Jun; 131(6):1710-7.

Detection of antibodies to Pseudomonas aeruginosa in serum and oral fluid from patients with cystic fibrosis. Weisner AM, Chart H, Bush A, **Davies JC**, Pitt TL. *J Med Microbiol*. 2007 May;56(Pt 5):670-4.

Safety of endobronchial biopsy in children with cystic fibrosis. Molina-Teran A, Hilliard TN, Saglani S, Haxby E, Scallan M, Bush A, **Davies JC.** *Pediatr Pulmonol*. 2006 Nov;41:1021-4.

Exploring the mechanisms of macrolides in cystic fibrosis. Equi AC, **Davies JC**, Painter H, Hyde S, Bush A, Geddes DM, Alton EW. *Respir Med*. 2006 Apr;100(4): 687-97.

Beta-defensin genomic copy number is not a modifier locus for cystic fibrosis. Hollox EJ, **Davies J,** Griesenbach U, Burgess J, Alton EW, Armour JA*. J Negat Results* Biomed. 2005 Dec 7;4:9.

Voriconazole therapy in children with cystic fibrosis. Hilliard T, Edwards S, Buchdahl R, Francis J, Rosenthal M, Balfour-Lynn I, Bush A, **Davies J.** *J Cyst Fibros*. 2005 Dec;4(4):215-20.

Tumour necrosis factor gene polymorphisms and childhood wheezing. Bilolikar H, Nam AR, Rosenthal M, **Davies JC,** Henderson DC, Balfour-Lynn IM. *Eur Respir J.* 2005 Oct;26(4):637-46.

Potential Difference Measurements in the Lower Airway of Children with and without Cystic Fibrosis. **Davies JC,** Davies M, McShane D, Smith S, Chadwick S, Jaffe A, Farley R, Collins L, Bush A, Scallon M, Pepper J, Geddes DM, Alton EW. *Am J Respir Crit Care Med.* 2005 May 1;171(9):1015-9.

Clinical improvement in cystic fibrosis following anti-tumourous chemotherapy. Eisen S, Painter H, Hyde SC, **Davies J**, Jaffe A. *Arch Dis Child*. 2004 Dec;89(12):1179-80.

Impaired pulmonary status in cystic fibrosis adults with 2 mutated *MBL-2* alleles. **JC Davies**, C Booth, M Johnson, N Shen, K Fidler, S Sharma, J Burgess, A Bush, DM Geddes, EWFW Alton, MW Turner, N Klein. *Eur Respir J.* 2004 Nov;24(5):798-804.

Normal nasal mucociliary clearance in CF children: evidence against a CFTR-related defect. D. McShane, **JC Davies**, T Wodehouse, A Bush, D Geddes, EWFW Alton. *Eur Respir J.* 2004 Jul;24(1)95-100.

Increased incidence and severity of the systemic inflammatory response syndrome in patients deficient in mannose-binding lectin. Fidler KJ, Wilson P, **Davies JC**, Turner MW, Peters MJ, Klein NJ. *Intensive Care Med*. 2004 Jul;30(7):1438-45.

Assays for human mannose-binding lectin. Turner MW, Johnson M, Booth C, Klein N, Rolland J, **Davies J**. *J Immunol Methods*. 2003 May 1;276(1-2):147-9.

A single round PCR method for genotyping human surfactant protein (SP)-A1, SP-A2 and SP-D gene alleles. P Pantelidis, AL Lagan**, JC Davies**, KI Welsh, RM du Bois. *Tissue Antigens* 2003 Apr;61(4):317-21.

Multi-resistant *Pseudomonas aeruginosa* in a paediatric CF centre: natural history and implications for segregation. Davies G, McShane D, **Davies J,** Bush A. *Pediatr Pulmonol* 2003 Apr;35(4):253-6.

Airway surface pH in subjects with cystic fibrosis. McShane D, **Davies JC**, Davies MG, Bush A, Geddes DM, Alton EWFW. *Eur Respir J. 2*003 Jan;21(1):37-42

Bone marrow stem cells do not repopulate the healthy upper respiratory tract. **Davies JC**, Potter M, Bush A, Rosenthal M, Geddes DM, Alton EWFW. *Pediatr Pulmonol.* 2002 Oct;34(4):251-6.

Airway function in infants newly diagnosed with cystic fibrosis. Ranganathan SC, Dezateux C, Bush A, Carr SB, Castle RA, Madge S, Price J, Stroobant J, Wade A, Wallis C, Stocks J and the **London Paediatric CF Collaboration**. *Lancet*. 2001 Dec 8;358(9297):1964-5.

Use of cough swabs in a cystic fibrosis clinic. Equi AC, Pike SE, **Davies J**, Bush A. *Arch Dis Child* 2001 Nov;85(5): 438-9.

Inflammation in cystic fibrosis airways: relationship to increased bacterial adherence. Scheid P, Kempster L, Griesenbach U, **Davies JC**, Dewar A, Webber PP, Colledge WH, Evans MJ, Geddes DM, Alton EW. *Eur Respir J* 2001 Jan;17(1):27-35

Differential binding of mannose-binding lectin to respiratory pathogens in cystic fibrosis. **Davies J,** Neth O, Alton E, Klein N, Turner M. *Lancet.* 2000 May 27;355(9218): 1885-6.

Cationic lipid-mediated *CFTR* gene transfer to the lungs and nose of CF subjects: A double-blind placebo-controlled trial. Alton EWFW, Stern M, Farley R, Jaffe A, Chadwick SL, Phillips J, **Davies J**, Smith SN *et al*. *Lancet.* 1999 Mar 20;353(9157):947-54.

Reduction of the adherence of *Pseudomonas aeruginosa* to native cystic fibrosis epithelium with anti-asialo GM1 antibody and neuraminidase inhibition. **Davies J,** Dewar A, Bush A, Pitt T, Gruenert D, Geddes DM, Alton EWFW. *Eur Respir. J* 1999 Mar;13(3):565-570.

Distal 10q trisomy syndrome with unusual cardiac and pulmonary abnormalities. **Davies J**, Jaffé A, Bush A. *J Med Genet*. 1998 Jan;35(1):72-4.

CFTR Gene Transfer reduces the binding of *Pseudomonas aeruginosa* to Cystic Fibrosis respiratory epithelial cells. **Davies J**, Stern M, Dewar A, Caplen NJ, Munkonge FM, Pitt T, Sorgi F, Huang L, Bush A, Geddes DM, Alton EWFW. *Am J Respir Cell Mol Biol.* 1997 Jun;16(6):657-63.

Retrospective Review of Effects of rhDNase in Children with CF. **Davies J,** Trindade M, Wallis C, Crawford O, Rosenthal M, Bush A. *Ped Pulmonol.* 1997 Apr;23(4):243-8.

Severe small airways disease resistant to medical treatment in a child with cystic fibrosis. **Davies J,** Rosenthal M, Bush A. *J R Soc Med.* 1996 Mar;89(3):172P-3P.

Hypogammaglobulinaemia associated with normal or increased IgM (the hyper IgM syndrome): a case series review. Banatvala N, **Davies J,** Kanariou M, Strobel S, Levinsky R, Morgan G. *Arch Dis Child.* 1994 Aug;71(2):150-2.

Preventing the scars of neonatal intensive care. **Davies J**, Gault D, Buchdahl R. *Arch Dis Child.* 1994 Jan;70(1):F50-1.

Review articles and editorials

Update on Clinical Outcomes of Highly Effective Modulator Therapy. Gifford AH, Taylor-Cousar JL, **Davies JC,** McNally P. *Clin Chest Med*. 2022;43(4):677-695.

Pseudomonas aeruginosa in the Cystic Fibrosis Lung. King J, Murphy R, **Davies JC**. *Adv Exp Med Biol*. 2022;1386:347-369.

New concepts in antimicrobial resistance in cystic fibrosis respiratory infections. Drevinek P, Canton R, Johansen HK, Hoffman L, Coenye T, Burgel P, **Davies JC.** *J Cyst Fibros*. 2022 Oct 18;S1569-1993(22)01387-X.

A systematic review of lung clearance index in non-cystic fibrosis, non-primary ciliary dyskinesia bronchiectasis. Hine C, Desai M, **Davies J**, Sapey E, Nagakumar P. *Respir Med*. 2022 Sep;201:106937.

Progress in Respiratory Gene Therapy. McLachlan G, Alton EWFW, Boyd AC, Clarke NK, **Davies JC**, Gill DR, Griesenbach U, Hickmott JW, Hyde SC, Miah KM, Molina CJ. *Hum Gene Ther.* 2022 Sep;33(17-18):893-912.

Going the Extra Mile: Why Clinical Research in Cystic Fibrosis Must Include Children. Dobra R, Bentley S, Edmondson C, Ovens M, Saunders C, Short C, Wilson G, **Davies JC**, Bush A. *Children (Basel)*. 2022 Jul 20;9(7):1080.

An Update on CFTR Modulators as New Therapies for Cystic Fibrosis. King JA, Nichols A, Bentley S, Carr SB, **Davies JC**. *Paediatr Drugs*. 2022 Jul;24(4):321-333.

Clinimetric analysis of outcome measures for airway clearance in people with cystic fibrosis: a systematic review. Stanford GE, Jones M, Charman SC, Bilton D, Usmani OS, **Davies JC**, Simmonds NJ. *Ther Adv Respir Dis*. 2022 Jan-Dec;16:17534666221122572.

Revisiting a diagnosis of cystic fibrosis - Uncertainties and considerations.Balfour-Lynn IM, Puckey M, Simmonds NJ, **Davies JC**. *Paediatr Respir Rev*. 2021 Dec 8:S1526-0542(21)00110-X.

COUNTERPOINT: In the Era of Cystic Fibrosis Transmembrane Regulator Protein Modulator Therapy, Are the Treatment Goals for Adults Now Different From Those for Children With Cystic Fibrosis? No. Thursfield RM, Shafi N, **Davies JC**. *Chest*. 2022 Jan;161(1):21-24.

The life rafts sailed; Now let's take stock and set the course ahead (Commentary). **Davies JC**. *J Cyst Fibros*. 2021 Dec;20 Suppl 3:29-30.

Time to get serious about the detection and monitoring of early lung disease in cystic fibrosis. Bayfield KJ, Douglas TA, Rosenow T, **Davies JC** et al. *Thorax*. 2021 Dec;76(12):1255-1265.

Model Systems to Study the Chronic, Polymicrobial Infections in Cystic Fibrosis: Current Approaches and Exploring Future Directions. O'Toole GA, Crabbé A, Kümmerli R, LiPuma JJ, Bomberger JM, **Davies JC** et al. *mBio*. 2021 Oct 26;12(5):e0176321.

Cystic fibrosis. Shteinberg M, Haq IJ, Polineni D, **Davies JC**. *Lancet*. 2021 Jun 5;397(10290):2195-2211.

Telemedicine and cystic fibrosis: Do we still need face-to-face clinics? E Dixon, K Dick, S Ollosson, D Jones, H Mattock, S Bentley, C Saunders, J Matthews, B Dobra, J King, C Edmondson, **J C Davies**. *Paediatr Respir Rev.* 2021 May 19;S1526-0542(21)00054-3.

Cystic Fibrosis Lung Disease Modifiers and Their Relevance in the New Era of Precision Medicine. Sepahzad A, Morris-Rosendahl DJ, **Davies JC**. *Genes (Basel)*. 2021 Apr 13;12(4):562.

Optimising equity of access: how should we allocate slots to the most competitive trials in Cystic Fibrosis (CF)? Dobra R, Davies G, Pike K, Strassle C, Allen L, Brendell R, Brownlee K, Carr SB, Simmonds NJ, **Davies JC**; London Network of Clinical Trials Accelerator Platform sites and affiliates. *J Cyst Fibros*. 2021 Apr 16:S1569-1993(21)00109-0.

Entering the era of highly effective modulator therapies. Dave K, Dobra R, Scott S, Saunders C, Matthews J, Simmonds NJ, **Davies JC**. *Pediatr Pulmonol*. 2021 Feb;56 Suppl 1:S79-S89.

JCF Year in Review. Flume PA, Castellani C, **Davies J**.*J Cyst Fibros*. 2021 Jan;20(1):1-2.

Updated guidance on the management of children with cystic fibrosis transmembrane conductance regulator-related metabolic syndrome/cystic fibrosis screen positive, inconclusive diagnosis (CRMS/CFSPID). Barben J, Castellani C, Munck A, **Davies JC** et al; Utility of lung clearance index in CF: What we know, what we don't know and musings on how to bridge the gap. Short C, Saunders C, **Davies JC**. *J Cyst Fibros*. 2020 Nov;19(6):852-855.

European CF Society Neonatal Screening Working Group (ECFS NSWG). *J Cyst Fibros*. 2020 Nov 27:S1569-1993(20)30909-7.

Monitoring early stage lung disease in cystic fibrosis. Nissenbaum C, Davies G, Horsley A, **Davies JC**. *Curr Opin Pulm Med*. 2020 Nov;26(6):671-678.

Investigating outcome measures for assessing airway clearance techniques in adults with cystic fibrosis: protocol of a single-centre randomised controlled crossover trial. Stanford G, **Davies JC** et al. *BMJ Open Respir Res*. 2020 Oct;7(1):e000694.

Gene Therapy for Respiratory Diseases: Progress and a Changing Context. Alton EWFW, Boyd AC, **Davies JC**, Gill DR, Griesenbach U, Harman TE, Hyde S, McLachlan G. Hum *Gene Ther*. 2020 Sep;31(17-18):911-916.

Restoration of exocrine pancreatic function in older children with cystic fibrosis on ivacaftor. Nichols AL, **Davies JC**, Jones D, Carr SB. *Paediatr Respir Rev*. 2020 Sep;35:99-102.

Horses for courses: Learning from functional tests of pulmonary health? Short C, Saunders C, **Davies JC**. *Pediatr Pulmonol*. 2020 Aug;55(8):1855-1858.

The coronavirus pandemic has forced rapid changes in care protocols for cystic fibrosis **Jane Davies**. *Nature*. 2020 Jul;583(7818):S15.

Cystic fibrosis drug trial design in the era of CFTR modulators associated with substantial clinical benefit: stakeholders' consensus view. De Boeck K, Lee T, Amaral M, Drevinek P, Elborn JS, Fajac I, Kerem E, **Davies JC**. *J Cyst Fibros*. 2020 Jun 8:S1569-1993(20)30162-4.

Building global development strategies for cf therapeutics during a transitional cftr modulator era. Mayer-Hamblett N, van Koningsbruggen-Rietschel S, Nichols DP, VanDevanter DR, **Davies JC** et al. *J Cyst Fibros*. 2020 Jun 7:S1569-1993(20)30161-2.

'Go for it, dream big, work hard and persist': A message to the next generation of CF leaders in recognition of International Women's Day 2020. Langawi MA, Byrnes C, **Davies JC** et al. *J Cyst Fibros*. 2020 Mar;19(2):184-193.

Selective Sampling of the Lower Airway in Children with Cystic Fibrosis: What Are We Missing? Turnbull A, Hughes D, **Davies J**. *Am J Respir Crit Care Med*. 2020 Mar 15;201(6):747-748.

The Lancet Respiratory Medicine Commission on the Future of Care of Cystic Fibrosis. Bell SC, Mall MA, Gutierrez H, Macek M, Madge S, **Davies JC** et al. *Lancet Respir Med.* 2019 Sep 27. pii: S2213-2600(19)30337-6.

Inconclusive diagnosis after a positive newborn bloodspot screening result for cystic fibrosis; clarification of the harmonised international definition. Southern KW, Barben J, Gartner S, Munck A, Castellani C, Mayell SJ, **Davies JC** et al. *J Cyst Fibros* 2019 Nov;18(6):778-780.

Speeding up access to new drugs for CF: Considerations for clinical trial design and delivery. **Davies JC**, Drevinek P, Elborn JS, Kerem E, Lee T; European CF Society (ECFS) Strategic Planning Task Force on ‘Speeding up access to new 4 drugs for CF’, Amaral MD, de Boeck K, Davies JC, Drevinek P, Elborn JS, Kerem E, Lee T. *J Cyst Fibros*. 2019 Sep;18(5):677-684.

Theranostics by testing CFTR modulators in patient-derived materials: The current status and a proposal for subjects with rare CFTR mutations. Amaral MD, de Boeck K; **ECFS Strategic**

Planning Task Force on 'Speeding up access to new drugs for CF'. *J Cyst Fibros*. 2019 Sep;18(5):685-692.

Trials and tribulations: The highs and lows of running cystic fibrosis drug studies. **Davies JC.** *Paediatr Respir Rev*. 2019 Mar 14. pii: S1526-0542(19)30025-9.

Chronic infection by controlling inflammation. Filloux A, **Davies JC**. *Nat Microbiol*. 2019 Mar;4(3):378-379.

"Fortunate are those who take the first steps"? The psychosocial impact of novel drug development. Dobra R, Madge S, Martin I, Weldon P, Simmonds N, **Davies JC**. *Paediatr Respir Rev*. 2019 Feb 27. pii: S1526-0542(19)30019-3.

Potentiators and Correctors in Paediatric Cystic Fibrosis Patients: A Narrative Review. Dobra R, Edmondson C, Hughes D, Martin I, **Davies JC**. *Paediatr Drugs*. 2018 Dec;20(6):555-566.

New anti-pseudomonal agents for cystic fibrosis- still needed in the era of small molecule CFTR modulators? **Davies JC**, Martin I. *Expert Opin Pharmacother*. 2018 Aug;19(12):1327-1336.

Predicting the Future of Cystic Fibrosis Lung Disease: Gene Expression Holds Some of the Answers. Edmondson C, **Davies JC**. *Ann Am Thorac Soc*. 2018 May;15(5):556-557.

Outdoor air pollution and cystic fibrosis. Brugha R, Edmondson C, **Davies JC**. *Paediatr Respir Rev.* 2018 Apr 11. pii: S1526-0542(18)30050-2.

Disease-modifying drug therapy in cystic fibrosis. Harman K, Dobra R, **Davies JC**. *Paediatr Respir Rev*. 2018 Mar;26:7-9.

Clinical trial research in focus: ensuring new cystic fibrosis drugs fulfil their potential. Elborn JS, **Davies J**. *Lancet Respir Med*. 2017 Sep;5(9):681-683.

Current and future therapies for Pseudomonas aeruginosa infection in patients with cystic fibrosis. Smith WD, Bardin E, Cameron L, Edmondson CL, Farrant KV, Martin I, Murphy RA, Soren O, Turnbull AR, Wierre-Gore N, Alton EW, Bundy JG, Bush A, Connett GJ, Faust SN, Filloux A, Freemont PS, Jones AL, Takats Z, Webb JS, Williams HD, **Davies JC**. *FEMS Microbiol Lett*. 2017 Aug 1;364(14).

Visualising early lung disease in CF: the emergence of MRI. **Davies JC.** *Thorax*. 2017 Aug;72(8):682.

Where are we with transformational therapies for patients with cystic fibrosis? De Boeck K, Davies JC. *Curr Opin Pharmacol*. 2017 Jun;34:70-75.

Developments in multiple breath washout testing in children with cystic fibrosis. Saunders C, Bayfield K, Irving S, Short C, Bush A, **Davies JC**. *Curr Med Res Opin*. 2017 Apr;33(4):613-620.

Current strategies for the long-term assessment, monitoring, and management of cystic fibrosis patients treated with CFTR modulator therapy. Elborn JS, **Davies J** et al. *J Cyst Fibros*. 2017 Jan;16(1):163-164.

Cystic fibrosis in 2016: considerable progress, but much more to do. **Davies JC**. *Lancet Respir Med*. 2016 Dec;4(12):943-945.

Cystic fibrosis gene therapy: a mutation-independent treatment. Griesenbach U, **Davies JC**, Alton E. *Curr Opin Pulm Med*. 2016 Nov;22(6):602-9.

Current and future treatment options for cystic fibrosis lung disease: latest evidence and clinical implications. Edmondson C, **Davies JC**. *Ther Adv Chronic Dis*. 2016 May;7(3):170-83.

Clinical trials of Novel Treatments for Cystic Fibrosis. Hippolyte S, Pabary R, Waller M, Jones A, Simmonds N, **Davies JC**. *Clinical Am J Respir Crit Care Med*. 2016;193:569-71.

Pseudomonas aeruginosa infection in cystic fibrosis: pathophysiological mechanisms and therapeutic approaches. Lund-Palau H, Turnbull AR, Bush A, Bardin E, Cameron L, Soren O, Wierre-Gore N, Alton EW, Bundy JG, Connett G, Faust SN, Filloux A, Freemont P, Jones A, Khoo V, Morales S, Murphy R, Pabary R, Simbo A, Schelenz S, Takats Z, Webb J, Williams HD, **Davies JC**. *Expert Rev Respir Med*. 2016 Jun;10(6):685-97.

New drug developments in the management of cystic fibrosis lung disease. Turnbull AR, **Davies JC**. *Expert Opin Pharmacother.* 2016 Jun;17(8):1103-12.

Multiple-Breath Washout as a Lung Function Test in Cystic Fibrosis. A Cystic Fibrosis Foundation Workshop Report. Subbarao P, Milla C, Aurora P, **Davies JC** et al. *Ann Am Thorac Soc*. 2015 Jun;12(6):932-9.

Recent advances in the management of cystic fibrosis. **Davies JC**, Ebdon AM, Orchard C. *Arch Dis Child.* 2014 Nov;99(11):1033-6.

Lung clearance index in primary ciliary dyskinesia and bronchiectasis. Irving SJ, **Davies JC** et al. *Am J Respir Crit Care Med*. 2014 May 1;189(9):1147-8.

Gene therapy in cystic fibrosis. Armstrong DK, Cunningham S, **Davies JC**, Alton EW. *Arch Dis Child*. 2014 May;99(5):465-8.

Lung clearance index: Evidence for use in clinical trials in cystic fibrosis. Kent L, Reix P, Innes JA, Zielen S, Le Bourgeois M, Braggion C, Lever S, Arets HG, Brownlee K, Bradley JM, Bayfield K, O'Neill K, Savi D, Bilton D, Lindblad A, **Davies JC**, Sermet I, De Boeck K; On behalf of the European Cystic Fibrosis Society Clinical Trial Network (ECFS-CTN) Standardisation Committee. *J Cyst Fibros*. 2014 Mar;13(2):123-38.

How to use: bacterial cultures in diagnosing lower respiratory tract infections in cystic fibrosis. Ahmed B, Bush A, **Davies JC**. *Arch Dis Child Educ Pract Ed.* 2014 Oct;99(5):181-7.

Cystic fibrosis: bridging the treatment gap in early childhood. **Davies JC**. *Lancet Respir Med*. 2013 Aug;1(6):433-4.

Genotype-specific small molecule therapy for cystic fibrosis. Thursfield RM, **Davies JC**. *Breathe* 2013; 9: 176-186.

A randomised, double-blind, placebo-controlled phase IIB clinical trial of repeated application of gene therapy in patients with cystic fibrosis. Alton EW, Boyd AC, Cheng SH, Cunningham S, **Davies JC** et al. *Thorax.* 2013 Nov;68(11):1075-7.

CFTR biomarkers: time for promotion to surrogate endpoint? De Boeck K, Kent L, **Davies J**, Derichs N, Amaral M, Rowe S, Middleton P, de Jonge H, Bronsveld I, Wilschanski M, Melotti P, Danner-Boucher I, Boerner S, Fajac I, Southern K, de Nooijer R, Bot A, de Rijke Y, de Wachter E, Leal T, Vermeulen F, J Hug M, Rault G, Nguyen-Khoa T, Barreto C, Proesmans M, Sermet-Gaudelus I; On behalf of the European Cystic Fibrosis Society Clinical Trial Network Standardisation Committee. *Eur Respir J.* 2013 Jan;41(1):203-16.

Thursfield RM, **Davies JC**. Cystic Fibrosis: therapies targeting specific gene defects. *Paediatr Respir Rev.* 2012 Dec;13(4):215-9.

Highlights of the North American Cystic Fibrosis Conference 2011. Pabary R, Thursfield R, **Davies JC**. *J R Soc Med*. 2012;105 Suppl 2:S9-S13

Basic science for the chest physician: Pseudomonas aeruginosa and the cystic fibrosis airway. Williams HD, **Davies JC**. *Thorax*. 2012 May;67:465-7

Gene therapy for the treatment of cystic fibrosis. Burney TJ, **Davies JC**. *Appl Clin Genet*. 2012 May 29;5:29-36.

Pseudomonas aeruginosa in cystic fibrosis: pathogenesis and new treatments. Brugha RE**,** **Davies JC**. *Br J Hosp Med (Lond)*. 2011 Nov;72(11):614-9.

Airway remodelling and its relationship to inflammation in cystic fibrosis. Regamey N, Jeffery PK, Alton EW, Bush A, **Davies JC**. *Thorax*. 2011 Jul; 66(7):624-9.

New clinical diagnostic procedures for cystic fibrosis in Europe. De Boeck K, Derichs N, Fajac I, de Jonge HR, Bronsveld I, Sermet I, Vermeulen F, Sheppard DN, Cuppens H, Hug M, Melotti P, Middleton PG, Wilschanski M; **ECFS Diagnostic Network Working Group**; EuroCareCF WP3 Group on CF diagnosis. *J Cyst Fibros.* 2011 Jun;10 Suppl 2:S53-66.

The journey of a thousand miles. Bush A, **Davies J**. *Eur Respir J.* 2011 Jan;37:8-9.

Design of gene therapy trials in CF patients.**Davies JC**, Alton EW. *Methods Mol Biol*. 2011;741:55-68.

Gene therapy for cystic fibrosis. **Davies JC**, Alton EW. *Proc Am Thorac Soc.* 2010 Nov;7(6):408-14.

Cystic fibrosis: to ion transport and beyond. Bush A, **Davies J**. *Eur Respir J.* 2010 Nov;36(5):991-2.

The future in paediatric respirology. Schmidt HJ, Bhandari V, Bhandari A, **Davies J** et al. *Respirology.* 2010 Jul;15(5):733-41.

Monitoring respiratory disease severity in cystic fibrosis. **Davies JC,** Alton EW. *Respir Care*. 2009 May;54(5):606-17.

Bugs, biofilms, and resistance in cystic fibrosis. **Davies JC**, Bilton D. *Respir Care*. 2009 May;54(5):628-40.

Lung clearance index in CF: a sensitive marker of lung disease severity. **Davies JC** et al. *Thorax*. 2008 Feb;63(2):96-7.

Cystic fibrosis. **Davies JC,** Alton EW, Bush A. *BMJ*. 2007 Dec 15;335(7632):1255-9.

Non! to non-steroidal anti-inflammatory therapy for inflammatory lung disease in cystic fibrosis (at least at the moment). Bush A, **Davies J.** *J Pediatr*. 2007 Sep;151(3):228-30.

Emerging and unusual gram-negative infections in cystic fibrosis. **Davies JC,** Rubin BK. *Semin Respir Crit Care Med*. 2007 Jun;28(3):312-21.

Biomarkers for cystic fibrosis: are we progressing? Alton EW, **Davies JC**, Geddes DM. *Am J Respir Crit Care Med.* 2007 Apr 15;175(8):750-1.

Computed tomography and cystic fibrosis: promises and problems. Aziz ZA, **Davies JC** et al. *Thorax*. 2007 Feb;62(2):181-6.

Novel antipseudomonal treatment approaches. **Davies JC**. *Arch Pediatr.* 2006 Oct;13 Suppl 1:S51-4.

Gene and cell therapy for cystic fibrosis. **Davies JC**. *Paediatr Respir Rev*. 2006;7 Suppl 1:S163-5.

New tests for cystic fibrosis. **Davies JC.** *Paediatr Respir Rev*. 2006;7 Suppl 1:S141-3.

Airway gene therapy. **Davies JC**, Alton EW. *Adv Genet.* 2005;54:291-314.

Cystic fibrosis modifier genes. **Davies JC,** Alton EWFW, Griesenbach U. *JRSM.* 2005;98 Suppl 45:47-54.

Early detection of lung disease in preschool children with cystic fibrosis. Bush A, **Davies J**. *Curr Opin Pulm Med.* 2005 Nov;11(6):534-8.

Modifier genes in cystic fibrosis. **Davies JC**, Griesenbach U, Alton E. *Pediatr Pulmonol.* 2005 May;39(5):383-91

Primum non nocere: does the current research publication system (or the lay press) harm our patients? **Davies J,** Bush A. *Am J Respir Crit Care Med* 2005 May 1;171(9): 937-8

Cystic fibrosis. **Davies JC**, Griesenbach U, Geddes DM, Alton EWFW. *Nature Encyclopaedia on line. In press*

Research applications of bronchoscopy. Payne D, **Davies JC**. *Paediatr Respir Revs.* 2003 Sep;4(3):230-6

Pseudomonas aeruginosa in Cystic Fibrosis: Pathogenesis and Persistence. **Davies J**. *Paediatr Respir Revs.* 2002 Jun;3(2):128-134.

New therapeutic approaches for cystic fibrosis lung disease. **Davies JC**. *J Royal Soc Med.* 2002;95 S41: 58-67.

Gene therapy for cystic fibrosis. **Davies JC,** Geddes DM, Alton EWFW. *J Gene Med.* 2001 Sep-Oct;3(5):409-417.

Prospects for gene therapy in lung disease. **Davies JC**, Geddes DM, Alton EWFW. *Curr Op Pharmacol.* 2001 Jun;1(3):272-7.

The role of the collectin system in pulmonary defence. **Davies JC**, Turner M, Klein N. *Paediatr Respir Revs.* 2001 Mar; 2(1):70-75.

Cystic fibrosis: an update of underlying aetiology and novel therapeutic approaches. **Davies J**, McShane D. *Curr Med Lit Paediatr.* 2001;14:53-57.

Prospects for gene therapy for cystic fibrosis. **Davies JC**, Geddes DM, Alton EW. *Mol Med Today*. 1998 Jul;4(7):292-9.

Infection in cystic fibrosis and congenital immune deficiency syndromes. **Davies J**, Bush A. *Curr Opin Infect Dis.* 1997;10:268-74.

New treatments for paediatric cystic fibrosis. **Davies J,** Hogg C, Rosenthal M. *Pediatr Tod.* 1996;4:81-84.

Importance of early inflammation in cystic fibrosis. **Davies J**, Bush A. *New Insights into Cystic Fibrosis.* 1995;3:5-8.

Letters/ images etc.

Nitrogen offset in N2 multiple washout method. Bayfield KJ, Alton E, Irving S, Bush A, **Davies JC**. *ERJ Open Res*. 2020 Mar 16;6(1):00043-2020.

Fair selection of participants in clinical trials: The challenge to push the envelope further. **Davies JC** et al. *J Cyst Fibros*. 2019 Sep;18(5):e48-e50.

Who and why; sharing our experiences of developing a standard operating procedure (SOP) to allocate screening slots for highly competitive cystic fibrosis trials. Dobra R, Scott S **Davies JC**, Simmonds NJ. *J Cyst Fibros* 2019 Sep;18(5):e45-e46

Cystic fibrosis presenting as acute upper airway obstruction. **Davies J,** Chaudry R, Larovere J, Hansell D, Dawson M, Abrahamson E. *Thorax*. 2006;61:92.

Measurement of tobramycin and gentamicin in saliva is not suitable for therapeutic drug monitoring of patients with cystic fibrosis. Spencer H, Kozlowska W, **Davies JC**, Webber P, Chadwick M, Kerr J, Makhecha S. *J Cyst Fibros*. 2005 Sep;4(3):209.

Edited books and contributed chapters

Textbooks

Hodson & Geddes' Cystic Fibrosis 5th Ed. Amaral, Bush, Davies, Ranganathan, Simmonds, Taylor-Cousar (2023; *In press*)

Current and Emerging Pharmaceutical Treatments for Cystic Fibrosis Lung Disease. Alton EWFW, **Davies JC**, Griesenbach U (Eds). Future Medicine, London, UK (2013).

An Atlas of Investigation and Management. Paediatric Respiratory Disease: Vol 2 Parenchymal Disease". Eds **Davies J** & Bush A. (Clinical Publishing) 2011.

An Atlas of Investigation and Management. Paediatric Respiratory Disease: Vol 1 Airways & Infection. Eds **Davies J** & Bush A. (Clinical Publishing) 2011.

Progress in Respiratory Reviews: Cystic Fibrosis in the 21st Century. Eds Alton, Bush, **Davies**, Griesenbach, Jaffe. Kaarger 2007Respiratory Research: Cystic Fibrosis in the 21st Century. Eds Alton, Bush, **Davies**, Griesenbach, Jaffe. Kaarger 2006.

Book Chapters

Molecular Therapies for Cystic Fibrosis. Davies G, Griesenbach U, Alton EWFW, **Davies JC.** In Disorders of the Respiratory Tract in Children. Elsevier 2016.

The Future. Basic Science: What will it deliver? **Davies JC,** Ratjen F. In Hodson & Geddes Cystic Fibrosis. Eds Bush, Bilton, Hodson. CRC Press 2016.

Outcome of Clinical Trials: Electricity, Induced Sputum and Breath. Waller M, Alton EWFW, **Davies JC**. In Hodson & Geddes Cystic Fibrosis. Eds Bush, Bilton, Hodson. CRC Press 2016.

Clinical Trials: What have we learned over the last 5 years? Elborn S, **Davies JC**, Bilton D. In Hodson & Geddes Cystic Fibrosis. Eds Bush, Bilton, Hodson. CRC Press 2016.

Modern Molecular Therapies for Respiratory Disease. Davies G, Alton EWFW, **Davies JC**. In Kendig & Chernick's Disorders of the Respiratory Tract in Children. Eds Wilmott, Boat, Bush, Chernick, Deterding, Ratjen. Elsevier 2012.

Cystic Fibrosis. **Davies J** & Bush A. In An Atlas of Investigation and Management. Paediatric Respiratory Disease: Vol 1 Airways & Infection. (Clinical Publishing) 2011.

Current and novel antimicrobial approaches. **Davies JC.** In Progress in Respiratory Research: Cystic Fibrosis in the 21st Century. Eds Alton, Bush, Davies, Griesenbach, Jaffe. Kaarger 2006.

Cystic Fibrosis Modifier Genes. Griesenbach U, Alton EWFW, **Davies JC**. In Progress in Respiratory Research: Cystic Fibrosis in the 21st Century. Eds Alton, Bush, Davies, Griesenbach, Jaffe. Kaarger 2006.

Respiratory Medicine. Davies J. In Essential Revision Notes in Paediatrics for the MRCPCH. Eds Beattie & Champion. PasTest 2006.

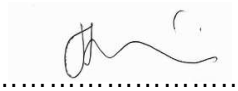
Acute infections producing upper airway obstruction. Balfour-Lynn IM, **Davies JC**. In Kendig's Disorders of the Respiratory tract in Childhood. Saunders 7th Edition 2006.

Airway Gene Therapy. **Davies JC**, Alton EWFW. In Non-viral vectors for Gene Therapy. Ed Huang, Hung & Wagner. Elsevier 2nd edition 2005.

Cystic Fibrosis in Childhood. Bush A and **Davies J**. In Respiratory Medicine. Ed Gibson, Geddes, Costabel, Sterk, Corrin. Saunders 3rd Ed 2003.

Gene Therapy Applications: Pulmonary. **Davies JC**, Geddes DM, Alton EWFW. In Pharmaceutical Gene Delivery Systems. Drugs and the Pharmaceutical Sciences Vol 131. Ed Rolland & Sullivan. Marcel Dekker 2003.

Respiratory. **Davies J**. In Essential Revision Notes in Paediatrics for the MRCPCH. Ed. Beattie and Champion. PasTest 1st Ed 2002.

Signed  …….

Date ……05/FEB/2022……..