Jane C Davies
Curriculum Vitae

Address 4 Orford Gardens, London, Twickenham TW1 4PL

D.O.B 9th November 1963

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

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GMC 3261765

Current Posts

2013 Onwards	Professor of Paediatric Respirology & Experimental Medicine Imperial College London
1999 Onwards	Honorary Consultant in Paediatric Respiratory Medicine Royal Brompton & Harefield NHS Foundation Trust, London

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, London
1989 – 1990	Senior House Officer in Paediatrics and Special Care, West Middlesex Hospital, London

Areas of Research Interest

Novel therapies and outcome measures for CF lung disease

Pathogenesis in CF:

- The innate defence system
- Airway remodelling and inflammation
- *Pseudomonas aeruginosa:* pathogenic mechanisms and non-invasive detection methods

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

BSc/ MBBS

Tamsin Lloyd[1] (2000-1)
Robert Shaw[1] (2001-2)
Arran Titley[1] (2002-3)
Louise Collins[1] (2003- 04)
Laura Benzonana[1] (2003-04)
Martha Truscott[1] (2004-05)
Fiona McLean[1] (2005-06)
Laura Wright[1] (2006-07)
Robert Fordham[1] (2007-08)
Sarah Moody[1] (2008-09)
Isis Redford[1] (2009-10)
Khayam Naderi[1] (2010; MBBS intercalated BSc)
Rajit Khosla[1] (2011; MBBS intercalated BSc)
Valerie Khoo[1] (2015; MBBS intercalated BSc)
Aaniya Ahmed[1] (2015; MBBS summer student)
Helena Lund Palau[1] (2015; MBBS Erasmus)
Laurance Pallant[1] (2016; MBBS intercalated BSc)
Nisal Weerakoon[1] (2016; MBBS intercalated BSc)
Ishani Seth[1] (2016; MBBS summer student)
Abigail Lark (2017; BSc summer student)

MSc

Yaqi Hu[1] (2016)

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)
Wynne Smith[1] (2015- ongoing)
Tom Semple[1] (2016- ongoing)
Rebecca Dobra[2] (2017 – 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)
Andrew Turnbull[1] (2014- writing up)
Natasha Wierre-Gore[2] (2015- ongoing)
Emmanuelle Bardin[2] (2015- ongoing)
Loren Cameron[2] (2015- ongoing)
Gemma Stanford[2] (2016- ongoing)
Claire Edmondson[1] (2016- ongoing)
Isaac Martin[1] (2016 – ongoing)
Dominic Hughes[1] (2017 – ongoing)
Ronan Murphy[1] (2017 – ongoing)

Post-doctoral fellows

Theresa Wodehouse (2003- 2005)
Nicholas Regamey (2005-2008)
Rossa Brugha (2016-ongoing; Academic Clinical Lecturer)

Grants Awarded

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

2017-2019 **Department of Health/ Wellcome Trust** **£2,773,098**

First-in-human trial of an optimised lentiviral vector for cystic fibrosis gene therapy

£81,312.58

2016-2019 **Cystic Fibrosis Trust** **(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.

DPFS Medical Research
2012 – 2013 **Council** **£1,243,400**
£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 Research Unit
2010 – 2011 **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

Committees

ECFS

- **Diagnostic Working Group Site Lead**
- **Standardisation Committee**
- **Task Force: Strategic Planning for faster access to new drugs**
- **Conference: Board (2016-ongoing); Scientific Committee (2004, 2012-15); President elect 2019**
- **Clinical Trials Network Principal Investigator**
- **Lung Clearance Index Core Facility Lead**

CF Trust

- Strategic Advisory Board (2013- ongoing)
- Clinical Trials Accelerator Platform Research & Scientific Oversight Committee
- CTAP London Network Lead Investigator

British Thoracic Society

- Science & Research Committee (2014- ongoing)
- Training & Education Committee (2014- ongoing)

Royal Brompton, Harefield and NHLI

- Research Ethics Committee (2007-2010)
- Clinical Research Oversight Committee (2015-ongoing)
- Respiratory Research Committee (2017- ongoing)

Other

- British Paediatric Respiratory Society: Research Committee Chair (2014- ongoing)
- British Lung Foundation: Paediatric Lung Conditions Research Advisory Board (2015- ongoing)
- Medicines for Children Research Network: Respiratory and CF CSG (2010- ongoing)
- UK CF Gene Therapy Consortium: Strategy Group and Clinical Lead (2003-ongoing)
- American Thoracic Society Pediatric International Relations Committee
- American Thoracic Society Conference Paediatric Assembly, 2007, 2008, 2009

Journal Editorial roles

- Associate Editor Thorax (2015- ongoing)
- Editorial Board Pediatric Allergy, Immunology and Pulmonology
- Series Editor: New Biology of the Airways, Paediatric Respiratory Reviews

Review/ referee activities

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

- Canadian CF Society
- Cystic Fibrosis in Australia
- German Federal Ministry of Education and Research Funding Initiative on Rare Disease Research Consortia
- Italian Research Foundation

Journal Publications:
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

Original Research

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. [Epub ahead of print]

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.

Tezacaftor-Ivacaftor in Residual-Function Heterozygotes with Cystic Fibrosis. Rowe, Steven M.; Daines, Cori; Ringshausen, Felix C.; Kerem, Eitan; Wilson, John; Tullis, Elizabeth; Nair, Nitin; Simard, Christopher; Han, Linda; Ingenito, Edward P.; Mckee, Charlotte; Lekstrom-Himes, Julie; **Davies, Jane C**. N Engl *J Med*. 2017 Nov 23;377(21):2024-2035.

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*. 2017 [Epub ahead of print].

The Immunomodulatory Drug Glatiramer Acetate is Also an Effective Antimicrobial Agent that Kills Gram-negative Bacteria. Stig Hill Christiansen, Ronan A. Murphy, Kristian Juul-Madsen, Marlene Fredborg, Michael Lykke Hvam, Esben Axelgaard, Sandra M. Skovdal, Rikke Louise Meyer, Uffe B. Skov Sørensen, Arne Möller, Jens Randel Nyengaard, Niels Nørskov-Lauritsen, Mikala Wang, Mihaela Gadjeva, Kenneth A. Howard, **Jane C. Davies**, Eskild Petersen, Thomas Vorup-Jensen. *Sci Rep*. 2017 Nov 15;7(1):15653.

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.

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**, Lee A Davies, Johanna F Dekkers, Ann Doherty, Sabrina Gea-Sorli, Deborah R Gill, Uta Griesenbach, Mamoru Hasegawa, Tracy E Higgins, Takashi Hironaka, Laura Hyndman, Gerry McLachlan, Makoto Inoue, Stephen C Hyde, J Alastair Innes, Toby M Maher, Caroline Moran, Cuixiang Meng, Michael C Paul-Smith, Ian A Pringle, Kamila M Pytel, Andrea Rodriguez-Martinez, Alexander C Schmidt, Barbara J Stevenson, Stephanie G Sumner-Jones, Richard Toshner, Shu Tsugumine, Marguerite W Wasowicz, Jie Zhu. *Thorax*. 2017 Feb; 72(2): 137–147.

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. 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;47:994-7.

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;4:107-15.

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**, Davies LA, Dewar MH, Doherty A, Donovan J, Dwyer NS, Elgmati HI, Featherstone RF, Gavino J, Gea-Sorli S, Geddes DM, Gibson JS, Gill DR, Greening AP, Griesenbach U, Hansell DM, Harman K, Higgins TE, Hodges SL, Hyde SC, Hyndman L, Innes JA, Jacob J, Jones N, Keogh BF, Limberis MP, Lloyd-Evans P, Maclean AW, Manvell MC, McCormick D, McGovern M, cLachlan G, Meng C, Montero MA, Milligan H, Moyce LJ, Murray GD, Nicholson AG, Osadolor T, Parra-Leiton J, Porteous DJ, Pringle IA, Punch EK, Pytel KM, Quittner AL, Rivellini G, Saunders CJ, Scheule RK, Sheard S, Simmonds NJ, Smith K, Smith SN, Soussi N, Soussi S, Spearing EJ, Stevenson BJ, Sumner-Jones SG, Turkkila M, Ureta RP, Waller MD, Wasowicz MY, Wilson JM, Wolstenholme-Hogg P; UK Cystic Fibrosis Gene Therapy Consortium. *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 1;192(11):1389-92.

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;373:220-31.

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. Lung clearance index in cystic fibrosis subjects treated for pulmonary exacerbations. *Eur Respir J*. 2015;46(4):1055-64.

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, 16;60(2):744-51.

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;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;46(6):1814-6.

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, 36:177-188

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;2:902-10.

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;44:1253-61.

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;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;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;34:10267-77.

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.

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