**Publications: (Recent only, not inclusive and in no particular order but grouped into areas of interest)**

**Exeter Team contribution to development of novel CFTR modulator therapy**

Middleton PG, Mall MA, Dřevínek P, et al

[Elexacaftor-Tezacaftor-Ivacaftor for Cystic Fibrosis with a Single Phe508del Allele.](https://www.ncbi.nlm.nih.gov/pubmed/31697873)

*N Engl J Med*. 2019 Nov 7;381(19):1809-1819.

Heijerman HGM, McKone EF, Downey DG et al

[Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: a double-blind, randomised, phase 3 trial.](https://www.ncbi.nlm.nih.gov/pubmed/31679946)

*Lancet*. 2019 Nov 23;394(10212):1940-1948.

Davies JC, Moskowitz SM, Brown C et al

[VX-659-Tezacaftor-Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles.](https://www.ncbi.nlm.nih.gov/pubmed/30334693)

*N Engl J Med.* 2018 Oct 25;379(17):1599-1611

Wainwright CE, Elborn JS, Ramsey BW et al

[Lumacaftor-Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508del CFTR.](https://www.ncbi.nlm.nih.gov/pubmed/25981758)

*N Engl J Med*. 2015 Jul 16;373(3):220-31.

**The role of Cardiopulmonary testing and aerobic fitness in CF .**

**This is an area the Exeter CF centre has worked extensively on in collaboration with Exeter University and been central in its development and very well respected in the CF community. We have many of you to thank for your contributions to this research!**

1. **Tomlinson, OW, Trott, J, Williams, CA, Withers NJ, Oades PJ.** Challenges in Im plementing Routine Cardiopulmonary Exercise Testing in Cystic Fibrosis Clinical Practice: a Single-Centre Review. SN Compr. Clin. Med. (2020). <https://doi.org/10.1007/> s42399-020-00239-7

2. **Tomlinson, O.W., Barker, A.R**., Fulford, J., Wilson, P., **Oades, P.J**. and Williams, C.A. Quantification of thigh muscle volume in children and adolescents using magnetic reso nance imaging. Eur J Sport Sci. 2020 Jan 12;1-10. doi: 10.1080/17461391.2019.1707292.

3. Analysis of Oxygen Uptake Efficiency Parameters in Young people with Cystic Fibrosis. **Owen W. Tomlinson**, Alan R. Barker, Lucy V. Chubbock, Daniel Stevens, Zoe L.Saynor, **Patrick J. Oades**, Craig A. Williams. Eur J Appl Physiol. 2018; 118(10): 2055–2063.

4. Oxygen Uptake Efficiency Slope is not a Valid Surrogate of Aerobic Fitness in Children with Cystic Fibrosis. CA Williams, **OW Tomlinson**, LV Chubbock, D Stevens, **ZL Saynor, PJ Oades,** AR Barker. Pediatr Pulmonol. 2018; 53(1):36-42.

5. Promotion of Exercise in the Management of Cystic Fibrosis – Summary of National Meetings. OW Tomlinson, J Shelley, S Denford, AR Barker, P**J Oades**, CA Williams. In Press: Eur J Pers Cent Healthcare 2018, Vol 6(2);196-203.

6. Measurement of V̇O2max in clinical groups is feasible and necessary. Craig Williams, **Zoe Saynor**, Alan Barker, **Patrick Oades, and Owen Tomlinson.** (Letter)

J Appl Physiol 2017; 123: 1017;doi:10.1152/japplphysiol.00538.

7. Scaling the Oxygen Uptake Efficiency Slope for Body Size in Cystic Fibrosis.

**Owen William Tomlinson,** Alan Robert Barker, **Patrick John Oades**, Craig Anthony Williams. Med Sci Sports Exerc. 2017; 49(10):1980-1986

8. Skeletal muscle oxidative capacity is/is not altered in patients with cystic fibrosis. Cross Talk article. Prof. Craig A. Williams, **Dr. Zoe L. Saynor, Owen Tomlinson, Dr. Patrick Oades,** Dr. Alan R. Barker. (Letter). J Physiol 2017 doi: 10.1113/JP272486

9. Exercise capacity following a percutaneous endoscopic gastrostomy in a young female with cystic fibrosis: A case report. **Owen W. Tomlinson**, Alan R. Barker, **Patrick J. Oades,** Craig A. Williams. Physiol Rep 2016; Aug; 4(16). pii: e12904.

10. Validity and reliability concerns associated with cardiopulmonary exercise testing young people with cystic fibrosis. Zoe L. Saynor, Dr. Alan R. Barker, **Dr. Patrick J. Oades**, Owen W. Tomlinson, Prof. Craig Anthony Williams. (Letter).  Respiration 2016 30;92(1):61-2.

11. Impaired Pulmonary V˙O2 Kinetics in Cystic Fibrosis Depend on Exercise Intensity. Zoe Saynor, Alan Barker, **Patrick Oades**, Craig Williams. Med Sci Sports Exerc. 2016; 48(11); 2090-2099.

  Feb 20 Challenges in Implementing Routine Cardiopulmonary Exercise Testing in Cystic Fibrosis Clinical Practice: a Single-Centre

<https://link.springer.com/article/10.1007/s42399-020-00239-7>

**T. Kent** , J**. Trott** , C.A. Williams , **P.J. Oades , N.J. Withers , O.W. Tomlinson** Ventilatory limitations to exercise in cystic fibrosis are dependent on lung function and equation selection. Abstract: *Journal of Cystic Fibrosis 2020;*Vol 19, S2.

**OW Tomlinson**, AR Barker, **J Trott,** **NJ Withers, PJ Oades**, CA Williams

Validity of prediction equations for evaluating aerobic fitness in cystic fibrosis.

Abstract: *Journal of Cystic Fibrosis 2020;*Vol 19, S2.

**O.W. Tomlinson,** A.R. Barker , **J. Trott** , P.J. Oades , **N.J. Withers**, C.A. Williams. Annual changes in aerobic fitness are biased between normative equations in children and adolescents with cystic fibrosis

Abstract: *Journal of Cystic Fibrosis 2020;*Vol 19, S2.

C.A. Williams, **O.W. Tomlinson**, **N.J. Withers** , **P.J. Oades**, A.R. Barker.

Differences in the ventilatory response to incremental exercise in people with cystic fibrosis according to age and sex.

Abstract: *Journal of Cystic Fibrosis 2020;*Vol 19, S2.

DJ Woolridge, **OW Tomlinson,** CL Bland, **PJ Oades, CD Sheldon**, **NJ Withers,**  CA Williams.

Predicting intravenous antibiotic usage with cardiopulmonary exercise testing in cystic fibrosis.

Abstract: *Journal of Cystic Fibrosis 2019;*Vol 18, S4.

C Williams, **O Tomlinson**, **N Withers, P Oades**, A Barker.

Sex and Age Differences in aerobic fitness in people with cystic fibrosis.

Abstract: *Journal of Cystic Fibrosis 2019;*Vol 18, S166.

**J Trott, O Tomlinson,** B Bowhay, C Williams, **N Withers,**  **P Oades.**

Reasons for non-compliance with cardiopulmonary exercise testing in cystic fibrosis.

Abstract: *Journal of Cystic Fibrosis 2018;*Vol 17, S101.

**CF microbiome and treatment**

Philip J Mitchelmore, Joanna Randall, Matthew J Bull, Karen A Moore, Paul A O’Neill, Konrad Paszkiewicz, Eshwar Mahenthiralingam, Chris J Scotton,

**Christopher D Sheldon**, **Nicholas J Withers**, Alan R Brown.

Molecular epidemiology of Pseudomonas aeruginosa in an unsegregated bronchiectasis cohort sharing hospital facilities with a cystic fibrosis cohort

*Thorax. 73. thoraxjnl-2016. 10.1136/thoraxjnl-2016-209889*

Coward A, Kenna DTD, Woodford N, Turton JF;  and **members of the UK CF Surveillance Working Group.**

[Structured surveillance of Achromobacter, Pandoraea and Ralstonia species from patients in England with cystic fibrosis.](https://www.ncbi.nlm.nih.gov/pubmed/31862307)

*J Cyst Fibros*. 2019 Dec 17. pii: S1569-1993(19)30967-1.

D. Smith, V. Duncan, T. Young, L. Katvars, D. Fraser-Pitt, **BJ Enderby**, **N.J. Withers**, C.D. Sheldon, **C. Auckland, P.J. Oades,** D. O'Neil

Novel approaches for CF associated respiratory fungal infections

Abstract: *Journal of Cystic Fibrosis 2016;*Vol 15, S1 PS43

L Anning, K Rider, K Pavel, **B Enderby, Oades P**, **Withers N**, **Sheldon C.**

Pathogen or Innocent Bystander? Review of Exophiala-Colonised Cystic Fibrosis Patients at a single UK Centre.

Abstract: *Paed Pulm* 2016, Vol 51 S45 P317

CF related Diabetes

Dr Lee Dobson completed his MD thesis in CF related Diabetes with great support from

Dr Chris Sheldon and the Exeter team - there are many research contributions that predate these if you are interested in Lees’s work but these are a taster.

***Sheldon CD*** *,* ***Dobson L***

**Insulin deficiency and diabetes related to cystic fibrosis.**

Journal of Cystic Fibrosis 2007 (May, p. 241)

**Dobson L**, Stride A, Bingham C, Elworthy S, Sheldon CD, Hattersley AT.

Microalbuminuria as a screening tool in cystic fibrosis related diabetes. *Pediatr Pulmonol* 2005 Feb; 39(2): 103-107.

**Dobson L**, Sheldon CD, Hattersley AT. Conventional measures underestimate glycaemia in Cystic Fibrosis (CF) patients. *Diabetic Medicine* 2004; 21(7): 691-6.

**Dobson L**, Sheldon CD, Hattersley AT. Understanding cystic-fibrosis-related diabetes: best thought of as insulin deficiency? *J R Soc Med* 2004; 97(Suppl.44): 1-10.

*Sheldon CD ,* ***Dobson L***

**Insulin deficiency and diabetes related to cystic fibrosis.**

Abstract: Journal of Cystic Fibrosis 2007 (May, p. 241)

Book chapter, **L Dobson** ’Cystic Fibrosis Related Diabetes’ – Cystic Fibrosis 3rd edition. Hodson, Geddes and Bush

General

**H Jones**, C Trealeaven, **L lane.** Parents experience of receiving a diagnosis of Cystic Fibrosis through the UK newborn hearing screening programme. Poster and Presentation European CF conference , Liverpool 2019

April 2017 and 2020.Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis, Third edition – Member of the working party ( **JTrott** and **Alice**)

**S. Whiteley** , J. Dalton , **N. Withers** , T. Shirazi . Early experiences of a bowel cancer screening programme in an adult cystic fibrosis centre

Abstract: *Journal of Cystic Fibrosis 2020;*Vol 19, S2.

Feb 2019 Is the management of Cystic Fibrosis improved by the use of online video calling to engage patients? A feasibility study using exercise as a model. https:// journals.sagepub.com/doi/pdf/10.1177/1357633X19828630

**H.J. Mortimer**, J. Shelley, **P.J. Oades, B.J. Enderby**, **N.J. Withers**, **C.D. Sheldon**

Complications of totally implantable venous access devices (TIVADs) in a CF centre.

Abstract: *Journal of Cystic Fibrosis 2016;* Vol 15, S1 P S103.

Barriers to compliance with and projected cost of nebulised treatment in Cystic Fibrosis: a CF centre perspective. **Trott J**, **Jones C, Rosie L, Green M, Shelley J, Enderby B**, Journal of CF 2016