

ECFS NEWSLETTER - Issue 52 - September 2016

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01. Letter from the President

Dear Friends,

I hope this finds you well and that you all have had a chance to enjoy some relaxing summer vacation.

For those of you who were able to participate in the June ECFS Conference in Basel, I am sure you will agree that it was a great success, and I would again like to thank the conference President Marc Chanson, and the Vice Presidents, Alexander Moeller and Juerg Barben for their contribution to the event. I would also like to thank the Cystic Fibrosis Association of Switzerland and all their considerable and enthusiastic support. My sincere thanks go to the Scientific Committee who produced an excellent programme, and to the speakers, moderators and presenters for their superb contribution.

The ECFS continued its tradition of awarding Travel Grants to young researchers based on the merit of their submitted abstracts, and we were delighted to welcome the 3 Young Investigator Awardees to the Closing Ceremony. All the award winners also received a 1 year free membership subscription to the ECFS.



Young Investigator Awards 2016: Lionelle Nkam (FR) Vilma Rautemaa (UK) Katie Bayfield (UK)

European Cystic Fibrosis Society Kastanieparken 7, 7470 Karup, Denmark Tel: +45 86 676260 Fax: +45 86 676290 Email: <u>info@ecfs.eu</u> Website: <u>www.ecfs.eu</u> To honour the enormous contribution of Prof.Gerd Döring, the ECFS has initiated a Gerd Döring Award in 2015 that is given annually to honour an exceptional early career young European scientist. This award includes a monetary grant of € 5000 to support research. This year, the Award was presented to Gillian Lavelle (IE) for her manuscript entitled "The basophil surface marker CD203c identifies Aspergillus species sensitisation in patients with cystic fibrosis". It describes for the first time the use of the novel basophil activation test for the rapid identification of Aspergillus fumigatus(Af)-sensitisation in people with CF, a state associated with poorer lung function. More information on the ECFS Awards:



Once again, speakers at this year's conference were asked to make their presentations (slides) available on the ECFS website (for members only) and these can be viewed by signing in under the "MY ECFS" tab in the top banner of our website <u>www.ecfs.eu</u>.

We have some changes in the ECFS Board and I would like to warmly welcome Isabelle Sermet and congratulate Margarida Amaral who is starting her second term. It's a pleasure to count them amongst the Board and we look forward to their input.

Again, I would like to thank Tanja Pressler, our departing Board member for her commitment and support during her term in office.

We are well into the planning for the Basic Science Conference which will be held in Albufeira in Portugal (29 March - 01 April 2017), and the annual conference which will be held 07-10 June 2017 in Seville, Spain. Programmes for both conferences will be available soon and we hope that you will take an active part in these events and consider submitting your best work for presentation and discussion. The abstract submission deadline for the conference in Seville will be 20 January 2017. More information about the conferences can be found on our website.

I would like to thank those members who participated in this year's Annual General Meeting in Basel, and you will find the minutes from the meeting included in this Newsletter.

As discussed at the AGM in Basel, our goal is to see 95% of patients with CF in every country to have 2 mutations identified by 2018. Please make use of the service and inform those around you about the ECFS CFTR gene sequencing service. More information later in the newsletter.

Finally, the EMA published a Concept paper on the need for revision of the guideline on the clinical development of medicinal products for the treatment of cystic fibrosis (CHMP/EWP/9147/08). This concept paper highlights items that have been identified and would need to be addressed in the revised guideline. The paper is open for comments (deadline of 31 October 2016).

As always, I would like to remind you to please feel free to send us articles that you would like to have included in forthcoming newsletters as this provides a great vehicle for communication.

Best wishes,

Kris De Boeck, ECFS President

02. Upcoming Events

- ECFS Board Meeting 26 October 2016 Orlando, Florida, USA
- 30th North American Cystic Fibrosis Conference 27-29 October 2016 Orlando, Florida, USA
- ECFS Winter Board Meeting 25 January 2017 Lisbon, Portugal
- ECFS CTN Steering Group Meeting 26-27 January 2017 Lisbon, Portugal
- ECFS Standards of Care Meeting 26 January 2017 Lisbon, Portugal
- ECFS Patient Registry Steering Group Meeting 27 January 2017 Lisbon, Portugal
- ECFS Diagnostic Network Working Group Meeting 16- 18 February 2017, Ljubljana, Slovenia
- 11th European Young Investigators Meeting 15-17 February 2017, Paris, France
- ECFS 14th Basic Science Conference 29 March 01 April 2017, Albufeira, Portugal
- ECFS Board Meeting 06 June 2017 Seville, Spain
- 40th European CF Conference 07-10 June 2017 Seville, Spain



03. Deadlines

- EMA Open consultation	31 October 2016
- Abstract submission ECFS Conference Seville	20 January 2017
- Abstract submission 14 th ECFS Basic Science conference	20 January 2017
- Nomination ECFS Award	15 February 2017
- Nomination Gerd Döring Award	15 February 2017
- Nomination ECFS Elections	30 March 2017

04. EMA Concept paper open for public consultation

The EMA would like to draw attention to the publication of the <u>Concept paper on the need for</u> revision of the guideline on the clinical development of medicinal products for the treatment of <u>cystic fibrosis (CHMP/EWP/9147/08)</u> open for public consultation.

This concept paper highlights items that have been identified and would need to be addressed in the revised guideline. We invite you to submit your comments, especially if you identify any overlooked issues.

The public consultation will run until **31 October 2016**. Comments should be provided using this <u>template</u> and sent to <u>respiratorydg@ema.europa.eu</u>.

Following this time period, EMA plans to release the draft Guideline for 6 months public consultation in October 2017, ending in April 2018.

05. CFTR gene sequencing service

As presented at the AGM in Basel, the ECFS ambitious goal is to see 95% of patients with CF in every country to have 2 mutations identified by 2018. The ECFS provides a service for highly parallel sequencing of the complete CFTR gene (including intronic and promoter regions) in patients with confirmed CF (maximum 5 patients per individual site) in whom a disease-causing mutation was not found on both CFTR genes. Please make use of the service and inform those around you about it! More information and request form can be found at https://www.ecfs.eu/ecfs_dnwg

06. Job postings

The BioSys PhD Programme in Life Sciences (http://biosys.campus.ciencias.ulisboa.pt) is an international programme for talented and highly motivated students of any nationality.

Research at the BioSys covers a range of topics with an interdisciplinary nature by applying integrative functional genomics and innovative research approaches. The programme supports its students in their scientific explorations, providing state-of-the-art equipment, expert scientific advisors, and extensive scientific and administrative support.

Applications for the 2016-2017 edition are open Click here (link is http://biosys.campus.ciencias.ulisboa.pt/) to have more details. Deadline for application 21 October 2016.

07. ECFS Education

The mission statement of the European Cystic Fibrosis Society is to improve survival and quality of life for people with CF by promoting high quality research, education and care. The ECFS has developed a number of educational activities so far: the ECFS annual conference, the Basic Science conference, the Journal of Cystic Fibrosis, development of guidelines and CF courses.

The ECFS is now working on the development of an educational website which will enhance the already existing activities and streamline all educational resources. In addition, the platform will allow for the development of compelling educational modules on different aspects of cystic fibrosis. Please contact us if you have a project or ideas for the educational website, we will be happy to discuss!



08. ECFS Board changes

We are happy to welcome Isabelle Sermet and Margarida Amaral to the ECFS Board. We would like to take this opportunity to warmly thank Tanja Pressler for her commitment through the years.



Thank you Tania!



Isabelle Sermet



Margarida Amaral

09. JCF Impact factor

The journal impact factors for 2015 have been announced by Thomson Reuters. The journal impact factor is a measure of how frequently papers from that particular journal are quoted. It is a pleasure to announce that the journal impact factor for the Journal of Cystic Fibrosis has increased this year from 3.475 to **3.853**.

This is the highest our impact factor has been since the journal started and reflects the high quality of papers published now in JCF.

10. JCF Research News

CF Research News offers lay version summaries of the most recent articles published in the Journal of Cystic Fibrosis. The aim of this new initiative is to bridge the gap between people with CF and the researchers investigating CF and how best to treat those with the condition. We aim to provide access to all scientific work published in the Journal of Cystic Fibrosis (JCF) to patients, parents, relatives, friends and caregivers of people with CF.

To access CF Research News click here

11. ECFS has got talent



At the Get-Together event that we will organise in Seville on the Friday 09 June 2017, we would like to put the spotlights on our ECFS members willing to show their talents. If you are a guitarist, a singer, a magician,... please get in touch with us, we would love to hear from you!

12. ECFS Tomorrow

The third edition of the EFCS Tomorrow in Basel was a great success.

Over the four days of conference, people from our multidisciplinary community researching and treating Cystic Fibrosis could meet at the ECFS Tomorrow Lounge to exchange ideas. Several interactive workshops and a debate were organised by Special Interest Groups and the feedback received indicated that all were enthusiastic about the initiative.

We also had our second edition of our ECFS Tomorrow Networking Event. During this relaxing evening friends and colleagues had the opportunity to meet new people and exchange experiences.

We also organised a quiz with a free registration to the Annual conference in Seville next year...and the winner is Rebecca Dobra, London, UK.

We are finalising the Seville programme, get ready for changes and look out for more information in the coming months!



13. Annual General Meeting Minutes

Minutes of the Annual General Meeting of the European Cystic Fibrosis Society

39th European Cystic Fibrosis Conference, Basel, Switzerland, 10th June 2016

Presiding Officer: Prof. Kris De Boeck

The meeting was opened at 18.30. Number of Participants: 40

President's Report

Prof. Kris De Boeck welcomed the ECFS members present and thanked them for their participation in the meeting. She hopes for higher number of participants at the next AGM in Seville.

She presented a slide listing the current ECFS Projects and Working Groups, and noted that reports on these activities had been provided ahead of the meeting and Coordinators of these projects would be ready to answer questions on their activities later on in the meeting.

Regarding ECFS conferences, she reported that about 2000 delegates had registered for the Basel conference and that 467 abstracts had been submitted. She took the opportunity to thank the Conference President and Conference Vice Presidents and all the other organizers for their contribution to a smooth preparation of the conference.

She went on to provide an overview of the ECFS activities over the last year, beginning with the Steering Committee meetings of the CTN and ECFSPR in January and the ECFS Diagnostic Network Working Group meeting in London in February. She mentioned the highly successful Basic Science conference held in Pisa, Italy, where a record number of attendees and abstract submissions were recorded.

The President then presented the programme of activities for 2016-2017 to include:

•	ECFS Autumn Meeting, Orlando CTN Steering Group Meeting, Lisbon	26 October 2016 26-27 January 2017
٠	ECFS Winter Board Meeting, Lisbon	25 January 2017
٠	Registry Steering Group Meeting, Lisbon	27 January 2017
٠	ECFS Diagnostic Network WG Meeting, Ljubljana	16-18 February 2017
٠	14 th ECFS Basic Science Conference, Albufeira	29 March - 1 April 2017
٠	Young Investigators Meeting, Paris	15-17 February 2017
•	40 th European CF Conference, Seville	7-10 June 2017

Prof. De Boeck presented the plans ahead for the society.

- Continuing CFTR gene sequencing service with a goal to see 95% of patients with CF in every country to have 2 mutations identified by 2018
- Defining research priorities : a strategic plan to maximize progress in CF therapies is being designed under the leadership of Jane Davies, London, UK
- Education: ECFS has ambitious projects and an educational portal is being developed under the leadership of Daniel Peckham, Leeds, UK
- Implementing mental health guidelines: the ECFS recently approved a new working group dedicated to mental health
- The ECFS website will be refurbished and will also include diary dates of meetings and a page dedicated to announcements for positions within CF.

- ECFS Book: the next ECFS book will be dedicated to infants and young children. Editors are sought for this new initiative and Prof De Boeck encouraged ECFS Members to volunteer.
- Further collaboration across continents

Prof. De Boeck presented some initiatives to further encourage participation of ECFS members to the activities of the Society.

- ECFS community bulletin board (Positions offered, jobs wanted, collaboration asked for, etc)
- Suggestion box for ECFS members at the ECFS conference (Suggestions for project opportunities, questions to ECFS board, nominations of candidates for future ECFS board positions, etc.)
- ECFS elections via e-voting also available during the ECFS conference
- Expansion of ECFS Tomorrow lounge activities
- Expansion of Special Interest Groups associated with ECFS

She thanked the departing ECFS Board Member Tania Pressler for her commitment over the years and ended her report by thanking the ECFS Membership, the National Patient Organisations, the Pharmaceutical Industry, and International collaborators for their continuing support of ECFS activities.

The audience was asked for any questions: None received.

Secretary's Report

Prof. Harm Tiddens reported the preliminary membership numbers for 2016 (May) were 925, an increase in comparison with the figures reported at the same time frame in 2015. One of the reasons for this was the introduction of conference registration and membership subscription available to all attendees.

In view of the healthy financial situation of the Society, he was happy to inform the membership that there would be no increase to the standard subscription fee in 2017; it will remain at Euro 120. There will be a continued lower rate for some categories of members who do not wish to receive the Journal of Cystic Fibrosis (JCF):

• Allied Health Professionals, PhD Students and Post Docs - 50 \notin (*)

• Retired members still actively engaged in an ECFS Project or Working Group - Free (*) (*) no JCF subscription associated with this rate

The discounted 3 years subscription (300 € instead of 360 €) introduced in 2015 will be offered as a membership possibility for the years 2017-2018-2019

He announced the introduction of a corporate membership rate for colleagues from the industry set at 220 \in and offering full membership benefits but no voting rights.

He thanked the departing Board members, Tacjana Pressler and Margarida Amaral for their invaluable input over the years.

The new Board members elected are: Margarida Amaral and Isabelle Sermet. Given the fact that the number of candidates corresponded to the number of positions available, no online secured voting was organised. The low number of candidates was disappointing and he encouraged the membership to stand for elections in the future.

The audience was asked for any questions: None received.

Treasurer's Report

Prof. Tania Pressler presented the audited ECFS Financial statement for the year 2015. The result of the year was very favourable, with a surplus of \notin 118,234 resulting in a positive balance for the Society's net assets of \notin 2,576,205 at 31st December 2015. Prof. Pressler informed the AGM that the full audited accounts will be posted on the website.

Update on Journal of Cystic Fibrosis

Dr. Scott Bell reported on developments of JCF since the start of his office. The acceptance rate is currently at 26%.

Based on the page budget, the number of paper published per annum should be in the region of 110 - 120.

There has been a steady increase of submitted articles (178 as of 6 June 2016) and a corresponding rise in the number of Science Direct downloads.

New initiatives and recent changes include:

- Ongoing renewal of Editorial Board (Carlo Castellani, Cliff Taggart)
- Streamline MS handling though finding 'reviewers' an ongoing challenge
- Editorials in each issue
- Bundled in themes
- "Invited Review" series (Endpoints)
- Highlights ("News" section)
- Lay Journal ("CF Research News")
- Best of at ECFC (Saturday 11 June) and NACFC
- Social media and the Journal launching Twitter

Report from ECFS CTN

Dr. Tim Lee presented a graph demonstrating the progression of studies conducted by the CTN since 2009.

He also presented the evolution of the CTN services including number of protocols reviewed and number of feasibility checks performed.

He reported on a recent survey of the CTN sites related to site capacity. Data will be used to apply for some targeted additional research infrastructure support from our European and North American partners

Report from ECFS Patient Registry

Dr. McKone reported on the activities of the ECFS Patient Registry (ECFSPR).

The ECFSPR is expanding its coverage with 28 countries participating and over 42,000 CF patients He reported that the ECFSPR aimed to publish the 2014 data report in the fall 2016. He presented a project for a benchmarking tool that will be developed allowing for

- Comparison between centres and centre-country
 - Comparison between centres and
 Comparison with previous years
 - Graphs in real time

He also reported on the recent development of a 2013 at a glance report in collaboration with CF Patient Organizations.

Finally, Dr. McKone thanked the project's sponsors and collaborators.

Report from ECFS Standards of Care

Pavel Drevinek reported on the progress of the Standards of Care project.

The best practice subgroup is reviewing and updating the 2014 Standards of Care. The goal is to get the document for review by the ECFS Board in March 2017 and ready for publication in May 2017. The Quality Management subgroup organized its second course in Basel. The group is developing a module in the ECFSTracker for benchmarking across countries/centres.

The centre framework subgroup is focusing on Standards of care in Eastern Europe. Analysis of a recent survey is underway.

Reports from ECFS Working Groups

Prior to the Annual General Meeting, members had been sent the progress reports from the ECFS Working Groups.

Dr Trudy Havermans presented the newly formed ECFS Mental Health working group. The working group is lead by Janice Abbott and Trudy Havermans and is working closely with the CFF Mental Health group.

Current Working Groups: Diagnostics Network Working Group Exercise Working Group Neonatal Screening Working Group Cystic Fibrosis Molecular & Cell Biology and Physiology Basic Science Working Group Mental Health Working Group The audience was asked for any questions: None received.

Special Interest Groups

Prof De Boeck reported on Special Interest Groups and their affiliation with ECFS: ECFS affiliated: ECFS Nurse Specialist Interest Group (ECFS NSiG) European Cystic Fibrosis Pharmacy Group (ECFPG) ECFS Psychosocial Working Group not ECFS affiliated: International Physiotherapy Group for Cystic Fibrosis European Cystic Fibrosis Nutrition Group

<u>Annual Conference 2017</u> A slide was shown presenting the 40th ECFS conference, to be held in Seville, Spain, 7-10 June 2017. Questions/Comments received: None

<u>Any Other Business</u> None

With no further items being raised, the meeting ended at 19.30.

14. 2016 Basel Conference Survey - Free Registration Winners

The ECFS would like to thank all those who took the time to answer the ECFS 2016 Survey after the conference in Basel. The answers and comments help us to keep on improving our conferences. As announced, three free registrations to the next ECFS Conference in Seville were selected from the respondents.

We congratulate the winners: Yulia Gorinova, Moscow, RU Roger Lundmark, Basel CH Elena Schneider, Sydney, AU

15. Current references in CF

Please scroll to next page for full list

CF Reference List

Adults & Adolescents

Baker JF., Putman MS., Herlyn K., Tillotson AP., Finkelstein JS., Merkel PA. Body composition, lung function, and prevalent and progressive bone deficits among adults with cystic fibrosis *Joint Bone Spine 2016; 83: 207 - 211*

Gutierrez CMA., Olivo MH., Moreno RMG. Dehydration in adult cystic fibrosis patients Archivos de Bronconeumologia 2016; 52: 398 - 399

Kazmerski TM., Borrero S., Tuchman LK., Weiner DJ., Pilewski JM., Orenstein DM., Miller E. Provider and Patient Attitudes Regarding Sexual Health in Young Women With Cystic Fibrosis Pediatrics 2016; 137: 6:e20154452

Mandaliya R., Hadjiliadis D., Cohen S. Gastroparesis Concurrent with Adult Cystic Fibrosis; Are They Related?

American Journal of Medicine 2016; 129: E21 - E22

Panchaud A., Di Paolo ER., Koutsokera A., Winterfeld U., Weisskopf E., Baud D., Sauty A., Csajka C. Safety of Drugs during Pregnancy and Breastfeeding in Cystic Fibrosis Patients *Respiration 2016; 91: 333 - 348*

Putman MS., Greenblatt LB., Sicilian L., Uluer A., Lapey A., Sawicki G., Gordon CM., Bouxsein ML., Finkelstein JS. Young adults with cystic fibrosis have altered trabecular microstructure by ITS-based morphological analysis Osteoporosis International 2016; 27: 2497 – 2505

Animal Model

Lavelle GM., White MM., Browne N., McElvaney NG., Reeves EP.

Animal Models of Cystic Fibrosis Pathology: Phenotypic Parallels and Divergences

Biomed Research International 2016; : ArNo: 5258727 **Meverholz DK.**

Lessons learned from the cystic fibrosis pig Theriogenology 2016; 86: 427 - 432

Vidovic D., Carlon M.S., da Cunha M.F., Dekkers J.F., Hollenhorst M.I., Bijvelds M.J., Ramalho A.S., Van den Haute C., Ferrante M., Baekelandt V., Janssens H.M., De Boeck K., Sermet-Gaudelus I., de Jonge H.R., Gijsbers R., Beekman J.M., Edelman A., et al

rAAV-CFTRAR Rescues the Cystic Fibrosis Phenotype in Human Intestinal Organoids and Cystic Fibrosis Mice American journal of respiratory and critical care medicine 2016; 193: 288 - 298

Yi YL., Sun XS., Gibson-Corley K., Xie WL., Liang B., He N., Tyler SR., Uc A., Philipson LH., Wang K., Hara M., Ode KL., Norris AW., Engelhardt JF.

A Transient Metabolic Recovery from Early Life Glucose Intolerance in Cystic Fibrosis Ferrets Occurs During Pancreatic Remodeling

Endocrinology 2016; 157: 1852 - 1865

Yi Y., Sun X., Gibson-Corley K., Xie W., Liang B., He N., Tyler S.R., Uc A., Philipson L.H., Wang K., Hara M., Ode K.L., Norris A.W., Engelhardt J.F.

A transient metabolic recovery from early life glucose intolerance in cystic fibrosis ferrets occurs during pancreatic remodeling

Endocrinology 2016; 157: 1852 - 1865

Antimicrobials

Atkinson CT., Tristram SG.

Antimicrobial resistance in cystic fibrosis isolates of Haemophilus influenzae British Journal of Biomedical Science 2016; 73: 87 - 89

Bandara HMHN., Herpin MJ., Kolacny D., Harb A., Romanovicz D., Smyth HDC. Incorporation of Farnesol Significantly Increases the Efficacy of

Liposomal Ciprofloxacin against Pseudomonas aeruginosa Biofilms in Vitro *Molecular Pharmaceutics 2016; 13: 2760 - 2770*

Barraud N., Kjelleberg S., Rice SA. Dispersal from Microbial Biofilms Microbiology Spectrum ; :

Batoni G., Maisetta G., Esin S.

Antimicrobial peptides and their interaction with biofilms of medically relevant bacteria *Biochimica et Biophysica Acta-Biomembranes 2016; 1858:* 1044 - 1060

Caballero JD., Pastor MD., Vindel A., Maiz L., Yague G., Salvador C., Cobo M., Morosini MI., del Campo R., Canton R.

Emergence of cfr-Mediated Linezolid Resistance in a Methicillin-Resistant Staphylococcus aureus Epidemic Clone Isolated from Patients with Cystic Fibrosis Antimicrobial Agents and Chemotherapy 2016; 60: 1878 - 1882

Cannavino CR., Mendes RE., Sader HS., Farrell DJ., Critchley IA., Biek D., Le J., Skochko SM., Jones RN., Bradley JS.

Evolution of ceftaroline-resistant MRSA in a child with cystic fibrosis following repeated antibiotic exposure *Pediatric Infectious Disease Journal 2016; 35: 813 - 815*

Cao B., Christophersen L., Kolpen M., Jensen PO., Sneppen K., Høiby N., Moser C., Sams T.

Diffusion Retardation by Binding of Tobramycin in an Alginate Biofilm Model

PLoS One 2016; 11: 4:e0153616

Caretti A., Torelli R., Perdoni F., Falleni M., Tosi D., Zulueta A., Casas J., Sanguinetti M., Ghidoni R., Borghi E., Signorelli P.

Inhibition of ceramide de novo synthesis by myriocin produces the double effect of reducing pathological inflammation and exerting antifungal activity against A-fumigatus airways infection

Biochimica et Biophysica Acta-General Subjects 2016; 1860: 1089 - 1097

d'Angelo I., Perfetto B., Costabile G., Ambrosini V., Caputo P., Miro A., Bianca RDD., Sorrentino R., Donnarumma G., Quaglia F., Ungaro F.

Large Porous Particles for Sustained Release of a Decoy Oligonucelotide and Poly(ethylenimine): Potential for Combined Therapy of Chronic Pseudomonas aeruginosa Lung Infections

Biomacromolecules 2016; 17: 1561 - 1571

Durham SH., Garza KB., Eiland LS.

Relationship between vancomycin dosage and serum trough vancomycin concentrations in pediatric patients with cystic fibrosis

American Journal of Health-system Pharmacy 2016; 73: 969 -974

Emiralioglu N., Yalcin E., Meral A., Sener B., Dogru D., Ozcelik U., Kiper N.

The success of the different eradication therapy regimens for Pseudomonas aeruginosa in cystic fibrosis

Journal of Clinical Pharmacy and Therapeutics 2016; 41: 419 - 423

Fischer DR., Namanny H., Zobell JT.

Follow-up survey of the utilization of anti-pseudomonal betalactam antibiotics at US cystic fibrosis centers *Pediatric Pulmonology 2016; 51: 668 - 669*

Forde E., Schutte A., Reeves E., Greene C., Humphreys H.,

Mall M., Fitzgerald-Hughes D., Devocelle M. Differential In Vitro and In Vivo Toxicities of Antimicrobial Peptide Prodrugs for Potential Use in Cystic Fibrosis Antimicrobial Agents and Chemotherapy 2016; 60: 2813 - 2821

Germoni LAP., Bremer PJ., Lamont IL.

The effect of alginate lyase on the gentamicin resistance of Pseudomonas aeruginosa in mucoid biofilms *Journal of Applied Microbiology 2016; 121: 126 - 135*

Horsley A., Jones AM., Lord R.

Antibiotic treatment for Burkholderia cepacia complex in people with cystic fibrosis experiencing a pulmonary exacerbation *Cochrane Database of Systematic Reviews* 2016; : 1:CD009529

Jain K., Wainwright C., Smyth AR.

Bronchoscopy-guided antimicrobial therapy for cystic fibrosis Cochrane Database of Systematic Reviews 2016; : 1:CD009530

Klare W., Das T., Ibugo A., Buckle E., Manefield M., Manos J.

Glutathione-Disrupted Biofilms of Clinical Pseudomonas aeruginosa Strains Exhibit an Enhanced Antibiotic Effect and a Novel Biofilm Transcriptome

Antimicrobial Agents and Chemotherapy 2016; 60: 4539 - 4551

Glutathione-Disrupted Biofilms of Clinical Pseudomonas aeruginosa Strains Exhibit an Enhanced Antibiotic Effect and a Novel Biofilm Transcriptome

Antimicrobial Agents and Chemotherapy 2016; 60: 4539 - 4551

Lenehan PJ., Schramm CM., Collins MS.

An evaluation strategy for potential QTc prolongation with chronic azithromycin therapy in cystic fibrosis Journal of Cystic Fibrosis 2016; 15: 192 - 195

Lo DKH., Hurley MN., Muhlebach MS., Smyth AR.

Interventions for the eradication of meticillin-resistant Staphylococcus aureus (MRSA) in people with cystic fibrosis Cochrane Database of Systematic Reviews 2015; : 2:CD009650

Mil-Homens D., Ferreira-Dias S., Fialho AM.

Fish oils against Burkholderia and Pseudomonas aeruginosa: in vitro efficacy and their therapeutic and prophylactic effects on infected Galleria mellonella larvae

Journal of Applied Microbiology 2016; 120: 1509 - 1519

Perez ALV., Schmidt-Malan SM., Kohner PC., Karau MJ., Greenwood-Quaintance KE., Patel R.

In vitro activity of ceftolozane/tazobactam against clinical isolates of Pseudomonas aeruginosa in the planktonic and biofilm states

Diagnostic Microbiology and Infectious Disease 2016; 85: 356 - 359

Pesavento G., Maggini V., Maida I., Lo Nostro A., Calonico C., Sassoli C., Perrin E., Fondi M., Mengoni A., Chiellini C., Vannacci A., Gallo E., Gori L., Bogani P., Bilia AR., Campana S., Ravenni N., Dolce D., Firenzuoli F., et al Essential Oil from Origanum vulgare Completely Inhibits the Growth of Multidrug-Resistant Cystic Fibrosis Pathogens Natural Product Communications 2016; 11: 861 - 864

Pompilio A., Crocetta V., Verginelli F., Di Bonaventura G. In vitro activity of levofloxacin against planktonic and biofilm Stenotrophomonas maltophilia lifestyles under conditions relevant to pulmonary infection in cystic fibrosis, and relationship with SmeDEF multidrug efflux pump expression *FEMS Microbiology Letters 2016; 363: 14:fnw145*

Qu L., She PF., Wang YX., Liu FX., Zhang D., Chen LH., Luo Z., Xu H., Qi Y., Wu Y.

Effects of norspermidine on Pseudomonas aeruginosa biofilm formation and eradication

Microbiology Open 2016; 5: 402 - 412

Samson C., Tamalet A., Thien HV., Taytard J., Perisson C., Nathan N., Clement A., Boelle PY., Corvol H.

Long-term effects of azithromycin in patients with cystic fibrosis

Respiratory Medicine 2016; 117: 1 - 6

Schneider EK., Azad MAK., Han ML., Zhou Q., Wang JP., Huang JX., Cooper MA., Doi Y., Baker MA., Bergen PJ., Muller MT., Li J., Velkov T.

An "Unlikely" Pair: The Antimicrobial Synergy of Polymyxin B in Combination with the Cystic Fibrosis Transmembrane Conductance Regulator Drugs KALYDECO and ORKAMBI ACS Infectious Diseases 2016; 2: 478 - 488

Turner RB., Elbarbry F., Biondo L.

Pharmacokinetics of once and twice daily dosing of intravenous tobramycin in paediatric patients with cystic fibrosis *Journal of Chemotherapy 2016; 28: 304 - 307*

van Velzen AJ., Bos AC., Touw DJ., Tiddens HAWM., Heijerman HGM., Janssens HM.

Pharmacokinetics and Tolerability of Once Daily Double Dose Tobramycin Inhalation in Cystic Fibrosis Using Controlled and Conventional Nebulization

Journal of Aerosol Medicine and Pulmonary Drug Delivery 2016; 29: 273 - 280

Wang WL., Yu JL., He Y., Wang ZL., Li F.

Ambroxol inhibits mucoid conversion of Pseudomonas aeruginosa and contributes to the bactericidal activity of ciprofloxacin against mucoid P-aeruginosa biofilms *APMIS 2016; 124: 611 - 618*

Wang ZM., Meenach SA.

Synthesis and Characterization of Nanocomposite Microparticles (nCmP) for the Treatment of Cystic Fibrosis-Related Infections

Pharmaceutical Research 2016; 33: 1862 - 1872

Wassermann T., Jorgensen KM., Ivanyshyn K., Bjarnsholt

T., Khademi SMH., Jelsbak L., Hoiby N., Ciofu O. The phenotypic evolution of Pseudomonas aeruginosa populations changes in the presence of subinhibitory concentrations of ciprofloxacin *Microbiology-SGM 2016; 162: 865 - 875*

Zhang HF., Nguyen MH., Clancy CJ., Joshi R., Zhao WC., Ensor C., Venkataramanan R., Shields RK. Pharmacokinetics of Posaconazole Suspension in Lung Transplant Patients with and without Cystic Fibrosis Antimicrobial Agents and Chemotherapy 2016; 60: 3558 – 3562

Cardiology

Coolen N., Gouya H., Kanaan R., Honore I., Chapron J., Hubert D., Legmann P., Dusser D., Burgel PR. Renin-associated hypertension after bronchial artery embolization in cystic fibrosis *Journal of Cystic Fibrosis 2016; 15: 213 - 215*

Rodriguez-Miguelez P., Thomas J., Seigler N., Crandall R., McKie KT., Forseen C., Harris RA.

Evidence of microvascular dysfunction in patients with cystic fibrosis

American Journal of Physiology-Heart and Circulatory Physiology 2016; 310: H1479 - H1485

Cell Biology

Bednarski C., Tomczak K., vom Hoevel B., Weber WM., Cathomen T.

Targeted Integration of a Super-Exon into the CFTR Locus Leads to Functional Correction of a Cystic Fibrosis Cell Line Model

PLoS One 2016; 11: 8:e0161072

Nyabam S., Wang Z., Thibault T., Oluseyi A., Basar R., Marshall L., Griffin M.

A novel regulatory role for tissue transglutaminase in epithelialmesenchymal transition in cystic fibrosis *Biochimica et Biophysica Acta-Molecular Cell Research 2016;*

1863: 2234 - 2244

Pincikova T., Svedin E., Domsgen E., Flodstrom-Tullberg M., Hjelte L.

Cystic fibrosis bronchial epithelial cells have impaired ability to activate vitamin D Acta Paediatrica 2016: 105: 851 - 853

Rymut SM., Kampman CM., Corey DA., Endres T., Cotton CU., Kelley TJ.

Ibuprofen regulation of microtubule dynamics in cystic fibrosis epithelial cells

American Journal of Physiology-Lung Cellular and Molecular Physiology 2016; 311: L317 - L327

CFTR

Atlante A., Favia M., Bobba A., Guerra L., Casavola V., Reshkin SJ.

Characterization of mitochondrial function in cells with impaired cystic fibrosis transmembrane conductance regulator (CFTR) function

Journal of Bioenergetics and Biomembranes 2016; 48: 197 - 210

Billeta A., Jia Y., Jensen TJ., Hou YX., Chang XB., Riordan JR., Hanrahan JW.

Potential sites of CFTR activation by tyrosine kinases Channels 2016; 10: 247 - 251

Borthwick L.A., Kerbiriou M., Taylor C.J., Cozza G., Lascu I., Postel E.H., Cassidy D., Trouv P., Mehta A., Robson L., Muimo R.

Role of interaction and nucleoside diphosphate kinase b in regulation of the cystic fibrosis transmembrane conductance regulator function by camp-dependent protein kinase a

PLoS One 2016; 11: 3:e0149097 -

Cantin AM.

Cystic Fibrosis Transmembrane Conductance Regulator Implications in Cystic Fibrosis and Chronic Obstructive Pulmonary Disease

Annals of the American Thoracic Society 2016; 13:

Carlile GW., Robert R., Matthes E., Yang Q., Solari R., Hatley R., Edge CM., Hanrahan JW., Andersen R., Thomas DY., Birault V.

Latonduine Analogs Restore F508del-Cystic Fibrosis Transmembrane Conductance Regulator Trafficking through the Modulation of Poly-ADP Ribose Polymerase 3 and Poly-ADP Ribose Polymerase 16 Activity

Molecular Pharmacology 2016; 90: 65 - 79

Cui GY., Khazanov N., Stauffer BB., Infield DT., Imhoff BR., Senderowitz H., McCarty NA.

Potentiators exert distinct effects on human, murine, and Xenopus CFTR

American Journal of Physiology-Lung Cellular and Molecular Physiology 2016; 311: L192 - L207

De Boeck K., Amaral MD.

Classification of CFTR mutation classes Reply Lancet Respiratory Medicine 2016; 4: E39

Dekkers JF., Berkers G., Kruisselbrink E., Vonk A., de Jonge HR., Janssens HM., Bronsveld I., de Graaf EAV., Nieuwenhuis EES., Houwen RHJ., Vleggaar FP., Escher JC., de Rijke YB., Majoor CJ., Heijerman HGM., de Winter-de Groot KM., et al

Characterizing responses to CFTR-modulating drugs using rectal organoids derived from subjects with cystic fibrosis *Science Translational Medicine 2016; 8: 344:344ra84*

Estacio SG., Martiniano HFMC., Faisca PFN.

Thermal unfolding simulations of NBD1 domain variants reveal structural motifs associated with the impaired folding of F508del-CFTR

Molecular Biosystems 2016; 12: 2834 - 2848

Farinha C.M., Swiatecka-Urban A., Brautigan D.L., Jordan P.

Regulatory crosstalk by protein kinases on CFTR trafficking and activity

Frontiers in Chemistry 2016; 4: RN1:1 -

Faure G., Bakouh N., Lourdel S., Odolczyk N., Premchandar A., Servel N., Hatton A., Ostrowski MK., Xu HJ., Saul FA., Moquereau C., Bitam S., Pranke I., Planelles G., Teulon J., Herrmann H., Roldan A., Zielenkiewicz P., Dadlez M., et al

Rattlesnake Phospholipase A(2) Increases CFTR-Chloride Channel Current and Corrects Delta F508CFTR Dysfunction: Impact in Cystic Fibrosis

Journal of Molecular Biology 2016; 428: 2898 - 2915

Flores AM., Casey SD., Felix CM., Phuan PW., Verkman AS., Levin MH.

Small-molecule CFTR activators increase tear secretion and prevent experimental dry eye disease *FASEB Journal 2016; 30: 1789 - 1797*

Gao XL., Hwang TC.

Spatial positioning of CFTR's pore-lining residues affirms an asymmetrical contribution of transmembrane segments to the anion permeation pathway

Journal of General Physiology 2016; 147: 407 - 422

Gianotti A., Capurro V., Scudieri P., Galietta LJV., Moran O., Zegarra-Moran O.

Pharmacological rescue of mutant CFTR protein improves the viscoelastic properties of CF mucus

Journal of Cystic Fibrosis 2016; 15: 295 - 301

Gottschalk LB., Vecchio-Pagan B., Sharma N., Han ST., Franca A., Wohler ES., Batista DAS., Goff LA., Cutting GR. Creation and characterization of an airway epithelial cell line for stable expression of CFTR variants *Journal of Cystic Fibrosis 2016; 15: 285 - 294*

Hollywood JA., Lee CM., Scallan MF., Harrison PT. Analysis of gene repair tracts from Cas9/gRNA double-stranded breaks in the human CFTR gene *Scientific Reports 2016; 6: ArNo: 32230*

Jun I., Cheng MH., Sim E., Jung J., Suh BL., Kim Y., Son H., Park K., Kim CH., Yoon JH., Whitcomb DC., Bahar I., Lee MG.

Pore dilatation increases the bicarbonate permeability of CFTR, ANO1 and glycine receptor anion channels *Journal of Physiology-London 2016; 594: 2929 - 2955*

Kim J., Noh SH., Piao H., Kim DH., Kim K., Cha JS., Chung WY., Cho HS., Kim JY., Lee MG. Monomerization and ER Relocalization of GRASP Is a

Requisite for Unconventional Secretion of CFTR Traffic 2016; 17: 733 - 753

Klein H., Abu-Arish A., Trinh NTN., Luo YS., Wiseman PW., Hanrahan JW., Brochiero E., Sauve R. Investigating CFTR and KCa3.1 Protein/Protein Interactions *PLoS One 2016; 11: 4:e0153665*

Kleme ML., Sane AT., Garofalo C., Levy E. Targeted CFTR gene disruption with zinc-finger nucleases in human intestinal epithelial cells induces oxidative stress and inflammation

International Journal of Biochemistry & Cell Biology 2016; 74: 84 - 94

Lobo MJ., Amaral MD., Zaccolo M., Farinha CM. EPAC1 activation by cAMP stabilizes CFTR at the membrane by promoting its interaction with NHERF1 *Journal of Cell Science 2016; 129: 2599 - 2612*

Lucarelli M., Bruno SM., Pierandrei S., Ferraguti G.,

Testino G., Truglio G., Strom R., Quattrucci S. The Impact on Genetic Testing of Mutational Patterns of CFTR Gene in Different Clinical Macrocategories of Cystic Fibrosis *Journal of Molecular Diagnostics 2016; 18: 554 - 565*

Mayer-Hamblett N., Boyle M., VanDevanter D.

Advancing clinical development pathways for new CFTR modulators in cystic fibrosis *Thorax 2016; 71: 454 - 461*

Miere C., Hewitson H., Wood V., Kadeva N., Cornwell G., Codognotto S., Stephenson E., Ilic D.

Generation of KCL021 research grade human embryonic stem cell line carrying a Delta F508 mutation in the CFTR gene *Stem Cell Research 2016; 16: 177 - 179*

Mihalyi C., Torocsik B., Csanady L.

Obligate coupling of CFTR pore opening to tight nucleotidebinding domain dimerization *eLife 2016; 5: ArNo: e18164*

Moisan S., Berlivet S., Ka C., Le Gac G., Dostie J., Ferec C.

Analysis of long-range interactions in primary human cells identifies cooperative CFTR regulatory elements *Nucleic Acids Research 2016; 44: 2564 - 2576*

Ong T., Ramsey BW.

New Therapeutic Approaches to Modulate and Correct Cystic Fibrosis Transmembrane Conductance Regulator *Pediatric Clinics of North America 2016; 63: 751 - +*

Palma AG., Galizia L., Kotsias BA., Marino GI.

CFTR channel in oocytes from Xenopus laevis and its regulation by xShroom1 protein *Pflugers Archiv-European Journal of Physiology 2016; 468:* 871 - 880

Qian F., Liu L., Liu ZZ., Lu CB.

The Pore Architecture of the Cystic Fibrosis Transmembrane Conductance Regulator Channel Revealed by Co-Mutation in Pore-Forming Transmembrane Regions *Physiological Research 2016; 65: 505 - 515*

Ramli N.S.K., Giribabu N., Muniandy S., Salleh N. Testosterone regulates levels of cystic fibrosis transmembrane regulator, adenylate cyclase, and cAMP in the seminal vesicles

of orchidectomized rats

Theriogenology 2016; 85: 238 - 246

Shah VS., Ernst S., Tang XX., Karp PH., Parker CP., Ostedgaard LS., Welsh MJ.

Relationships among CFTR expression, HCO3- secretion, and host defense may inform gene- and cell-based cystic fibrosis therapies

Proceedings of the National Academy of Sciences of the United States 2016; 113: 5382 - 5387

Sherwood CL., Boitano S.

Airway epithelial cell exposure to distinct e-cigarette liquid flavorings reveals toxicity thresholds and activation of CFTR by the chocolate flavouring 2,5-dimethypyrazine *Respiratory Research 2016; 17: ArNo: 57*

Solomon GM., Raju SV., Dransfield MT., Rowe SM.

Therapeutic Approaches to Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in Chronic Bronchitis

Annals of the American Thoracic Society 2016; 13:

Sorio C., Montresor A., Bolomini-Vittori M., Caldrer S., Rossi B., Dusi S., Angiari S., Johansson JE., Vezzalini M., Leal T., Calcaterra E., Assael BM., Melotti P., Laudanna C. Mutations of Cystic Fibrosis Transmembrane Conductance Regulator Gene Cause a Monocyte-Selective Adhesion

Deficiency American Journal of Respiratory and Critical Care Medicine

2016; 193: 1123 - 1133

Sosnay PR., Raraigh KS., Gibson RL.

Molecular Genetics of Cystic Fibrosis Transmembrane Conductance Regulator: Genotype and Phenotype Pediatric Clinics of North America 2016; 63: 585 -

Tosco A., De Gregorio F., Esposito S., De Stefano D., Sana I., Ferrari E., Sepe A., Salvadori L., Buonpensiero P., Di Pasqua A., Grassia R., Leone CA., Guido S., De Rosa G., Lusa S., Bona G., Stoll G., Maiuri MC., Mehta A., et al A novel treatment of cystic fibrosis acting on-target: cysteamine plus epigallocatechin gallate for the autophagy-dependent rescue of class II-mutated CFTR

Cell Death and Differentiation 2016; 23: 1380 - 1393

van der Mark VA., de Jonge HR., Chang JC., Ho-Mok KS., Duijst S., Vidovic D., Carlon MS., Elferink RPJO., Paulusma CC.

The phospholipid flippase ATP8B1 mediates apical localization of the cystic fibrosis transmembrane regulator *Biochimica et Biophysica Acta-Molecular Cell Research 2016; 1863: 2280 - 2288*

Veit G., Oliver K., Apaja PM., Perdomo D., Bidaud-Meynard A., Lin ST., Guo JY., Icyuz M., Sorscher EJ., Hartman JL., Lukacs GL.

Ribosomal Stalk Protein Silencing Partially Corrects the Delta F508- CFTR Functional Expression Defect *PLoS Biology 2016; 14: 5:e1002462*

Vetter AJ., Karamyshev AL., Patrick AE., Hudson H., Thomas PJ.

N-Alpha-Acetyltransferases and Regulation of CFTR Expression

PLoS One 2016; 11: 5:e0155430

Vetter A.J., Karamyshev A.L., Patrick A.E., Hudson H., Thomas P.J.

N-alpha-acetyltransferases and regulation of CFTR expression PLoS One 2016; 11: 5:e0155430 -

Watson MJ., Lee SL., Marklew AJ., Gilmore RC., Gentzsch M., Sassano MF., Gray MA., Tarran R.

The Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Uses its C-Terminus to Regulate the A2B Adenosine Receptor

Scientific Reports 2016; 6: ArNo: 27390

Xue RQ., Gu H., Qiu YM., Guo Y., Korteweg C., Huang J., Gu J.

Expression of Cystic Fibrosis Transmembrane Conductance Regulator in Ganglia of Human Gastrointestinal Tract Scientific Reports 2016; 6: ArNo: 30926

Yan C., Lang Q., Liao HJ., Jiang X., Ming Y., Sun HQ., Xu WM.

CFTR Deletion in Mouse Testis Induces VDAC1 Mediated Inflammatory Pathway Critical for Spermatogenesis *PLoS One 2016; 11: 8:e0158994*

Yang R., Kerschner JL., Gosalia N., Neems D., Gorsic LK., Safi A., Crawford GE., Kosak ST., Leir SH., Harris A. Differential contribution of cis-regulatory elements to higher order chromatin structure and expression of the CFTR locus *Nucleic Acids Research 2016; 44: 3082 - 3094*

Yu YC., Sohma Y., Hwang TC.

On the mechanism of gating defects caused by the R117H mutation in cystic fibrosis transmembrane conductance regulator *Journal of Physiology-London 2016; 594: 3227 - 3244*

Zwick M., Esposito C., Hellstern M., Seelig A.

How Phosphorylation and ATPase Activity Regulate Anion Flux though the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)

Journal of Biological Chemistry 2016; 291: 14483 – 14498

Clinical

Adler FR., Liou TG.

The Dynamics of Disease Progression in Cystic Fibrosis *PLoS One 2016; 11: 6:e0156752*

Bourke SJ., Booth Z., Doe S., Anderson A., Rice S.,

Gascoigne A., Quibell R. A service evaluation of an integrated model of palliative care of cystic fibrosis

Palliative Medicine 2016; 30: 698 - 702

Davis SD., Ratjen F., Brumback LC., Johnson RC., Filbrun AG., Kerby GS., Panitch HB., Donaldson SH., Rosenfeld M. Infant lung function tests as endpoints in the ISIS multicenter clinical trial in cystic fibrosis

Journal of Cystic Fibrosis 2016; 15: 386 - 391

De Boeck K., Bulteel V., Fajac I.

Disease-specific clinical trials networks: the example of cystic fibrosis

European Journal of Pediatrics 2016; 175: 817 - 824

Lee AL., Rawlings S., Bennett KA., Armstrong D.

Pain and its clinical associations in individuals with cystic fibrosis: A systematic review *Chronic Respiratory Disease 2016; 13: 102 - 117*

Chronic Respiratory Disease 2010; 15: 102 - 11

Lewis D.

Helping patients with cystic fibrosis live longer Journal of Family Practice 2016; 65: 188 - 194

Martin C., Hamard C., Kanaan R., Boussaud V., Grenet D.,

Abely M., Hubert D., Munck A., Lemonnier L., Burgel PR. Causes of death in French cystic fibrosis patients: The need for improvement in transplantation referral strategies! *Journal of Cystic Fibrosis 2016; 15: 204 - 212*

McIntosh ID.

Health Human Resources Guidelines: Minimum Staffing Standards and Role Descriptions for Canadian Cystic Fibrosis Healthcare Teams

Canadian Respiratory Journal 2016; : ArNo: 6369704

Miller R., Bourke S., Immanuel A., Metcalfe S. Subtotal Esophagectomy for Carcinoma in a Patient with Cystic Fibrosis

A & E Case Reports 2016; 6: 394 - 396

Silva AM., Descalco A., Salgueiro M., Pereira L., Barreto C., Bandeira T., Ferreira R.

Respiratory sleep disturbance in children and adolescents with cystic fibrosis

Revista Portuguesa de Pneumologia 2016; 22: 202 - 208

Steyger P.S., Garinis A.C.

Hearing Loss Risk Factors for Cystic Fibrosis Patients Hearing Journal 2015; 69: 8 - 9

Tremblay F., Chapdelaine H., Lavoie A., Berthiaume Y., Sabbah L., Bernstein SC., Silviet-Carricart M. Toxic epidermal necrolysis in a patient with cystic fibrosis Journal of Allergy and Clinical Immunology-in Practice 2016; 4: 526 – 528

Databases & Registries

Knapp EA., Fink AK., Goss CH., Sewall A., Ostrenga J., Dowd C., Elbert A., Petren KM., Marshall BC.

The Cystic Fibrosis Foundation Patient Registry Design and Methods of a National Observational Disease Registry Annals of the American Thoracic Society 2016; 13: 1173 – 1179

Diabetes

Armaghanian N., Brand-Miller JC., Markovic TP., Steinbeck KS.

Hypoglycaemia in cystic fibrosis in the absence of diabetes: A systematic review

Journal of Cystic Fibrosis 2016; 15: 274 - 284

Belle-van Meerkerk G., de Valk HW., Stam-Slob MC., van Berkhout FT., Zanen P., van de Graaf EA.

Cystic Fibrosis-Related Diabetes with strict glycaemic control is not associated with frequent intravenous antibiotics use for pulmonary infections

Diabetes Research and Clinical Practice 2016; 116: 230 - 236

Boudreau V., Coriati A., Desjardins K., Rabasa-Lhoret R. Glycated hemoglobin cannot yet be proposed as a screening tool for cystic fibrosis related diabetes

Journal of Cystic Fibrosis 2016; 15: 258 - 260

Burgess JC., Bridges N., Banya W., Gyi KM., Hodson ME., Bilton D., Simmonds NJ.

HbA1c as a screening tool for cystic fibrosis related diabetes Journal of Cystic Fibrosis 2016; 15: 251 - 257

Burgess JC., Bridges N., Winston B., Gyi KM., Hodson ME., Bilton D., Simmonds NJ. HbA1c as a screening tool for cystic fibrosis related diabetes

Response Journal of Cystic Fibrosis 2016; 15: 265 - 266

Coriati A., Dubois CL., Phaneuf M., Mailhot M., Lavoie A., Berthiaume Y., Rabasa-Lhoret R.

Relationship between vitamin D levels and glucose tolerance in an adult population with cystic fibrosis *Diabetes & Metabolism 2016; 42: 135 - 138*

Coriati A., Ziai S., Lavoie A., Berthiaume Y., Rabasa-Lhoret R.

The 1-h oral glucose tolerance test glucose and insulin values are associated with markers of clinical deterioration in cystic fibrosis

Acta Diabetologica 2016; 53: 359 - 366

Koivula FNM., McClenaghan NH., Harper AGS., Kelly C. Islet-intrinsic effects of CFTR mutation *Diabetologia 2016; 59: 1350 - 1355*

Montanini L., Cirillo F., Smerieri A., Pisi G., Giardino I., d'Apolito M., Spaggiari C., Bernasconi S., Amarri S., Street ME.

HMGB1 Is Increased by CFTR Loss of Function, Is Lowered by Insulin, and Increases In Vivo at Onset of CFRD Journal of Clinical Endocrinology & Metabolism 2016; 101: 1274 - 1281

Onady GM., Stolfi A.

Insulin and oral agents for managing cystic fibrosis-related diabetes

Cochrane Database of Systematic Reviews 2016; : 4:CD004730

Prentice B., Hameed S., Verge CF., Ooi CY., Jaffe A., Widger J.

Diagnosing cystic fibrosis-related diabetes: current methods and challenges

Expert Review of Respiratory Medicine 2016; 10: 799 - 811

Schnyder MA., Benden C., Schmid C. HbA1c: An effective screening tool for cystic fibrosis related diabetes?

Journal of Cystic Fibrosis 2016; 15: 261 - 262

Widger J., Hameed S., Ooi CY., Verge C. Using HbA1c as a screening tool for Cystic Fibrosis Related Diabetes

Journal of Cystic Fibrosis 2016; 15: 263 – 264

Diagnosis

Rosenfeld M., Sontag MK., Ren CL.

Cystic Fibrosis Diagnosis and Newborn Screening Pediatric Clinics of North America 2016; 63: 599 - +

Endocrinology

Blackman SM., Tangpricha V. Endocrine Disorders in Cystic Fibrosis Pediatric Clinics of North America 2016; 63: 699 - +

Lee SY., Chesdachai S., Lee MJ., He XM., Tangpricha V., Braverman LE.

Thyroid Function in Patients with Cystic Fibrosis: No Longer a Concern? Thyroid 2016; 26: 875 – 879

Epidemiology

Silva A., Amorim A., Azevedo P., Lopes C., Gamboa F. Cystic fibrosis - characterization of the adult population in Portugal *Revista Portuguesa de Pneumologia 2016; 22: 141 - 145*

Stewart C., Pepper MS. Cystic fibrosis on the African continent Genetics in Medicine 2016; 18: 653 - 662

Vahedi L., Jabarpoor-Bonyadi M., Ghojazadeh M., Hazrati H., Rafeev M.

Association between Outcomes and Demographic Factors in an Azeri Turkish Population With Cystic Fibrosis: A Cross-Sectional Study in Iran From 20 01 through 2014 *Iranian Red Crescent Medical Journal 2016; 18: 4:e29615*

Exercise

Del Coso J., Lara B., Salinero JJ., Areces F., Ruiz-Vicente D., Gallo-Salazar C., Abian-Vicen J., Cacabelos R.

CFTR genotype-related body water and electrolyte balance during a marathon

Scandinavian Journal of Medicine & Science in Sports 2016; 26: 1036 -1044

Doeleman WR., Takken T., Bronsveld I., Hulzebos EHJ.

Relationship between lung function and Modified Shuttle Test performance in adult patients with cystic fibrosis: a crosssectional, retrospective study

Physiotherapy 2016; 102: 184 - 188

Hebestreit H., Arets HGM., Aurora P., Boas S., Cerny F., Hulzebos EHJ., Karila C., Lands LC., Lowman JD., Swisher A., Urquhart DS.

Validity and Reliability Concerns Associated with Cardiopulmonary Exercise Testing Young People with Cystic Fibrosis Reply

Respiration 2016; 92: 63 - 64

Kriemler S., Radtke T., Christen G., Kerstan-Huber M., Hebestreit H.

Short-Term Effect of Different Physical Exercises and Physiotherapy Combinations on Sputum Expectoration, Oxygen Saturation, and Lung Function in Young Patients with Cystic Fibrosis

Lung 2016; 194: 659 - 664

Malamud AL., Ricard PEH.

Feasibility of the Six-Minute Walk Test for Patients who have Cystic Fibrosis, are Ambulatory, and Require Mechanical Ventilation before Lung Transplantation *Physical Therapy 2016; 96: 1468 - 1476*

Saynor ZL., Barker AR., Oades PJ., Tomlinson OW., Williams CA.

Validity and Reliability Concerns Associated with Cardiopulmonary Exercise Testing Young People with Cystic Fibrosis

Respiration 2016; 92: 61 - 62

Vallier JM., Rouissi M., Mely L., Gruet M.

Physiological Responses of the Modified Shuttle Test in Adults With Cystic Fibrosis Journal of Cardiopulmonary Rehabilitation and Prevention

2016; 36: 288 - 292

Van Iterson EH., Wheatley CM., Baker SE., Morgan WJ., Snyder EM.

The relationship between cardiac hemodynamics and exercise tolerance in cystic fibrosis *Heart & Lung 2016; 45: 283 – 290*

Gastroenterology

Brecelj J., Zidar N., Jeruc J., Orel R.

Morphological and Functional Assessment of Oesophageal Mucosa Integrity in Children With Cystic Fibrosis Journal of Pediatric Gastroenterology and Nutrition 2016; 62: 757 - 764

Corral JE., Dye CW., Mascarenhas MR., Barkin JS., Salathe M., Moshiree B.

Is Gastroparesis Found More Frequently in Patients with Cystic Fibrosis? A Systematic Review Scientifica 2016; : ArNo: 2918139

Demeyer S., De Boeck K., Witters P., Cosaert K. Beyond pancreatic insufficiency and liver disease in cystic fibrosis

European Journal of Pediatrics 2016; 175: 881 - 894

Hatziagorou E., Kampouras A., Sidiropoulou M., Markou A., Anastasiou A., Tsanakas J.

Pancreatic Cystosis in Two Adolescents with Cystic Fibrosis Case Reports in Pediatrics 2016; : ArNo: 5321785

Howlett C., Ronan NJ., NiChroinin M., Mullane D., Plant BJ.

Partial restoration of pancreatic function in a child with cystic fibrosis

Lancet Respiratory Medicine 2016; 4: E21 - E22

Ooi CY., Jeyaruban C., Lau J., Katz T., Matson A., Bell SC., Adams SE., Krishnan U.

High ambient temperature and risk of intestinal obstruction in cystic fibrosis

Journal of Paediatrics and Child Health 2016; 52: 430 - 435 Sathe MN., Freeman AJ.

Gastrointestinal, Pancreatic, and Hepatobiliary Manifestations of Cystic Fibrosis

Pediatric Clinics of North America 2016; 63: 679 - +

Gene Therapy

Karda R., Buckley S.M., Waddington S.N.

Gene Therapy with Adeno-associated Virus for Cystic Fibrosis American Journal of Respiratory and Critical Care Medicine 2016; 193: 234 - 236

Paul-Smith MC., Bell RV., Alton WE., Alton EWFW., Griesenbach U.

Gene therapy for cystic fibrosis: recent progress and current aims

Expert Opinion on Orphan Drugs 2016; 4: 649-658

General Review

VanDevanter DR., Kahle JS., O'Sullivan AK., Sikirica S., Hodgkins PS.

Cystic fibrosis in young children: A review of disease manifestation, progression, and response to early treatment *Journal of Cystic Fibrosis 2016; 15: 147 – 157*

Genetics

Diana A., Polizzi AM., Santostasi T., Ratclif L., Pantaleo MG., Leonetti G., Iusco DR., Gallo C., Conese M., Manca A. The novel complex allele [A238V;F508del] of the CFTR gene: clinical phenotype and possible implications for cystic fibrosis etiological therapies

Journal of Human Genetics 2016; 61: 473 - 481

Gajbhiye R., Kadam K., Khole A., Gaikwad A., Kadam S., Shah R., Kumaraswamy R., Khole V.

Cystic fibrosis transmembrane conductance regulator (CFTR) gene abnormalities in Indian males with congenital bilateral absence of vas deferens & renal anomalies *Indian Journal of Medical Research 2016*, 143: 616 - 623

Marson FAL., Bertuzzo CS., Ribeiro JD.

Classification of CFTR mutation classes Lancet Respiratory Medicine 2016; 4: E37 - E38

Salinas DB., Sosnay PR., Azen C., Young S., Raraigh KS., Keens TG., Kharrazi M.

Benign and Deleterious Cystic Fibrosis Transmembrane Conductance Regulator Mutations Identified by Sequencing in Positive Cystic Fibrosis Newborn Screen Children from California

PLoS One 2016; 11: 5:e0155624

Shahin WA., Mehaney DA., El-Falaki MM.

Mutation spectrum of Egyptian children with cystic fibrosis Springerplus 2016; 5: ArNo: 686

Stanke F., Tűmmler B. Classification of CFTR mutation classes Lancet Respiratory Medicine 2016; 4: E36

Temurhan S., Tamay Z., Gurkan H., Akgul S., Ozceker D., Kekik C., Cagatay P., Aydin F., Guler N. The Effect of TGFB1 and CD14 Gene Polymorphisms on the Clinical Findings of Cystic Fibrosis in Turkish Patients

International Journal of Human Genetics 2016; 16: 40 - 47

Zomer-van Ommen DD., Vijftigschild LAW., Kruisselbrink E., Vonk AM., Dekkers JF., Janssens HM., de Winter-de Groot KM., van der Ent CK., Beekman JM.

Limited premature termination codon suppression by readthrough agents in cystic fibrosis intestinal organoids Journal of Cystic Fibrosis 2016; 15: 158 – 162

Growth & Development

Kelly A., Schall J., Stallings VA., Zemel BS.

Trabecular and cortical bone deficits are present in children and adolescents with cystic fibrosis *Bone 2016; 90: 7 - 14*

Knepper C., Ellemunter H., Eder J., Niedermayr K., Haerter B., Hofer P., Scholl-Burgi S., Muller T., Heinz-Erian P.

Low sodium status in cystic fibrosis-as assessed by calculating fractional Na+ excretion-is associated with decreased growth parameters

Journal of Cystic Fibrosis 2016; 15: 400 - 405

Naydenova-Stoeva B.

Markers of bone metabolism in patients with cystic fibrosis Journal of IMAB 2015; 21: 901 - 907

Stein L., Pacht C., Junge S., Kaeding TS., Kuck M., Maassen N., Wittke T., Shushakov V.

Skeletal Muscle Function in Young Patients with Cystic Fibrosis Pediatric Exercise Science 2016; 28: 364 - 373

Tejero S., Cejudo P., Quintana-Gallego E., Sanudo B., Oliva-Pascual-Vaca A.

The role of daily physical activity and nutritional status on bone turnover in cystic fibrosis: a cross-sectional study *Brazilian Journal of Physical Therapy 2016; 20: 206 - 212*

Zhang Z., LindstromM.J., Farrell P.M., Lai H.J.

Pubertal height growth and adult height in cystic fibrosis after newborn screening

Pediatrics 2016; 137: 5:e20152907 –

Immunology & Inflammation

Averna M., Bavestrello M., Cresta F., Pedrazzi M., De Tullio R., Minicucci L., Sparatore B., Salamino F., Pontremoli S., Melloni E.

Abnormal activation of calpain and protein kinase C alpha promotes a constitutive release of matrix metalloproteinase 9 in peripheral blood mononuclear cells from cystic fibrosis patients *Archives of Biochemistry and Biophysics 2016; 604: 103 - 112*

Bostik V., Prasil P., Plisek S., Kracmarova R., Kosina P., Salavec M., Sleha R., Chlibek R., Bostik P.

Breakthrough Varicella Zoster Virus Infection in an Immunized Child with Cystic Fibrosis

Pediatric Infectious Disease Journal 2016; 35: 595 - 596

Chhuon C., Pranke I., Borot F., Tondelier D., Lipecka J., Fritsch J., Chanson M., Edelman A., Ollero M., Guerrera IC.

Changes in lipid raft proteome upon TNF-alpha stimulation of cystic fibrosis cells.

Journal of Proteomics ; 145: 246-253

Chillappagari S., Müller C., Mahavadi P., Guenther A., Nährlich L., Rosenblum J., Rubin B.K., Henke M.O. A small molecule neutrophil elastase inhibitor, KRP-109, inhibits cystic fibrosis mucin degradation *Journal of Cystic Fibrosis 2016; 15: 325 - 331*

Debyser G., Mesuere B., Clement L., Van de Weygaert J., Van Hecke P., Duytschaever G., Aerts M., Dawyndt P., De Boeck K., Vandamme P., Devreese B.

Faecal proteomics: A tool to investigate dysbiosis and inflammation in patients with cystic fibrosis Journal of Cystic Fibrosis 2016; 15: 242 - 250

Laval J., Ralhan A., Hartl D. Neutrophils in cystic fibrosis *Biological Chemistry 2016; 397: 485 - 496*

Le Moigne V., Gaillard JL., Herrmann JL. Vaccine strategies against cystic fibrosis pathogens Human Vaccines & Immunotherapeutics 2016; 12: 751 - 756

Meijer L., Nelson DJ., Riazanski V., Gabdoulkhakova AG., Hery-Arnaud G., Le Berre R., Loaec N., Oumata N., Galons H., Nowak E., Gueganton L., Dorothee G., Prochazkova M., Hall B., Kulkarni AB., Gray RD., Rossi AG., Witko-Sarsat V.

Modulating Innate and Adaptive Immunity by (R)-Roscovitine: Potential Therapeutic Opportunity in Cystic Fibrosis Journal of Innate Immunity 2016; 8: 330 - 349

Poghosyan A., Patel JK., Clifford RL., Knox AJ.

Epigenetic dysregulation of interleukin 8 (CXCL8) hypersecretion in cystic fibrosis airway epithelial cells *Biochemical and Biophysical Research Communications 2016;* 476: 431 - 437

Pradenas GA., Ross BN., Torres AG.

Burkholderia cepacia Complex Vaccines: Where Do We Go from here?

Vaccines 2016; 4: 2:10

Prentice B., McKay K., Selvadurai H., Robinson PD., Abel F., Fitzgerald DA.

Question 6: Is there a role for Mannose-Binding Lectin measurement in Cystic Fibrosis management? Paediatric Respiratory Reviews 2016; 19: 46 – 48

Liver Disease

Drzymala-Czyz S., Jonczyk-Potoczna K., Lisowska A., Stajgis M., Walkowiak J.

Supplementation of ursodeoxycholic acid improves fat digestion and absorption in cystic fibrosis patients with mild liver involvement

European Journal of Gastroenterology & Hepatology 2016; 28: 645 - 649

Lemaitre C., Dominique S., Billoud E., Eliezer M., Montialoux H., Quillard M., Riachi G., Koning E., Morisse-Pradier H., Savoye G., Savoye-Collet C., Goria O.

Relevance of 3D Cholangiography and Transient Elastography to Assess Cystic Fibrosis-Associated Liver Disease? *Canadian Respiratory Journal 2016; : ArNo: 4592702*

Palaniappan S.K., Than N.N., Moe S., van Mourik I., Thein A.W.

Interventions for managing advanced liver disease in cystic fibrosis

Cochrane Database of Systematic Reviews 2016; 2016: CD012056 -

Pratico AD., Pratico ER., Rotolo N., Salafia S., Franzonello C., Leonardi S.

Isolated liver disease in a patient with a CFTR genotype F508del/12TG- 5T and 470MV: A new face of an old disease Annals of Hepatology 2015; 14: 933 - 936

Stonebraker JR., Ooi CY., Pace RG., Corvol H., Knowles MR., Durie PR., Ling SC.

Features of Severe Liver Disease With Portal Hypertension in Patients With Cystic Fibrosis

Clinical Gastroenterology and Hepatology 2016; 14: 1207 - +

Microbiology

Ahmed MI., Mukherjee S. Treatment for chronic Staphylococcus aureus pulmonary infection in people with cystic fibrosis Cochrane Database of Systematic Reviews 2015; : 3:CD011581

Al-Momani H., Perry A., Stewart CJ., Jones R., Krishnan A., Robertson AG., Bourke S., Doe S., Cummings SP., Anderson A., Forrest T., Griffin SM., Brodlie M., Pearson J., Ward C.

Microbiological profiles of sputum and gastric juice aspirates in Cystic Fibrosis patients

Scientific Reports 2016; 6: ArNo: 26985

Albrecht C., Ringshausen F., Ott S., Wagner D.,

Rademacher J., Schneider M., Welte T., Pletz M.W. Should all adult cystic fibrosis patients with repeated nontuberculous mycobacteria cultures receive specific treatment? A 10-year case-control study

European Respiratory Journal 2016; 47: 1575 - 1577

Alshraiedeh NH., Higginbotham S., Flynn PB., Alkawareek MY., Tunney MM., Gorman SP., Graham WG., Gilmore BF.

Eradication and phenotypic tolerance of Burkholderia cenocepacia biofilms exposed to atmospheric pressure nonthermal plasma

International Journal of Antimicrobial Agents 2016; 47: 446 - 450

Ambrose M., Malley RC., Warren SJC., Beggs SA., Swallow OFE., McEwan B., Stock D., Roddam LF.

Pandoraea pnomenusa Isolated from an Australian Patient with Cystic Fibrosis

Frontiers in Microbiology 2016; 7: ArNo: 692

Aubert DF., Xu H., Yang JL., Shi XY., Gao WQ., Li L., Bisaro F., Chen S., Valvano MA., Shao F.

A Burkholderia Type VI Effector Deamidates Rho GTPases to Activate the Pyrin Inflammasome and Trigger Inflammation *Cell Host & Microbe 2016; 19: 664 - 674*

Bacci G., Paganin P., Lopez L., Vanni C., Dalmastri C., Cantale C., Daddiego L., Perrotta G., Dolce D., Morelli P., Tuccio V., De Alessandri A., Fiscarelli EV., Taccetti G., Lucidi V., Bevivino A., Mengoni A.

Pyrosequencing Unveils Cystic Fibrosis Lung Microbiome Differences Associated with a Severe Lung Function Decline *PLoS One 2016; 11: 6:e0156807*

Bernier SP., Workentine ML., Li X., Magarvey NA., O'Toole GA., Surette MG.

Cyanide Toxicity to Burkholderia cenocepacia Is Modulated by Polymicrobial Communities and Environmental Factors *Frontiers in Microbiology 2016; 7: ArNo: 725*

Betran A., Villuendas MC., Rezusta A., Pereira J., Revillo M.J., Rodriguez-Nava V.

Clinical significance, antimicrobial susceptibility and molecular identification of Nocardia species isolated from children with cystic fibrosis

Brazilian Journal of Microbiology 2016; 47: 531 - 535

Caballero JD., del Campo R., Royuela A., Sole A., Maiz L., Olveira C., Quintana-Gallego E., de Gracia J., Cobo M., de la Pedrosa EGG., Oliver A., Canton R., Sole A., Cortell I., Asensio O., Garcia G., Martinez MT., Cols M., Salcedo Bronchopulmonary infection-colonization patterns in Spanish cystic fibrosis patients: Results from a national multicenter study

Journal of Cystic Fibrosis 2016; 15: 357 - 365

Caverly LJ., Carmody LA., Haig SJ., Kotlarz N., Kalikin LM., Raskin L., LiPuma JJ.

Culture-Independent Identification of Nontuberculous Mycobacteria in Cystic Fibrosis Respiratory Samples *PLoS One 2016; 11: 4:e0153876*

Chotirmall SH.

Candida albicans in cystic fibrosis: "Opening statements presented, let the trial begin" *Pediatric Pulmonology 2016; 51: 445 - 446*

Cigana C., Melotti P., Baldan R., Pedretti E., Pintani E., Iansa P., De Fino I., Favari F., Bergamini G., Tridello G., Cirillo DM., Assael BM., Bragonzi A. Genotypic and phenotypic relatedness of Pseudomonas

aeruginosa isolates among the major cystic fibrosis patient cohort in Italy BMC Microbiology 2016; 16: ArNo: 142

Cools P., Ho E., Vranckx K., Schelstraete P., Wurth B., Franckx H., Ieven G., Van Simaey L., Van Daele S., Verhulst S., De Baets F., Vaneechoutte M. Epidemic Achromobacter xylosoxidans strain among Belgian cystic fibrosis patients and review of literature

BMC Microbiology 2016; 16: ArNo: 122

Cuthbertson L., Rogers GB., Walker AW., Oliver A., Green LE., Daniels TWV., Carroll MP., Parkhill J., Bruce KD., van der Gast CJ.

Respiratory microbiota resistance and resilience to pulmonary exacerbation and subsequent antimicrobial intervention ISME Journal 2016; 10: 1081 - 1091

Depluverez S., Devos S., Devreese B.

The Role of Bacterial Secretion Systems in the Virulence of Gram-Negative Airway Pathogens Associated with Cystic Fibrosis

Frontiers in Microbiology 2016; 7: ArNo: 1336

Dingemans J., Ghequire MGK., Craggs M., De Mot R., Cornelis P.

Identification and functional analysis of a bacteriocin, pyocin S6, with ribonuclease activity from a Pseudomonas aeruginosa cystic fibrosis clinical isolate

Microbiology Open 2016; 5: 413 - 423

Downes KJ., Abulebda K., Siracusa C., Moore R., Staat MA., Poynter SE.

Non-typeable Haemophilus influenzae purulent pericarditis in a child with cystic fibrosis

Pediatrics International 2016; 58: 607 - 609

Eltringham I., Pickering J., Gough H., Preece CL., Perry JD.

Comparison of Mycobacterial Growth Indicator Tube with Culture on RGM Selective Agar for Detection of Mycobacteria in Sputum Samples from Patients with Cystic Fibrosis Journal of Clinical Microbiology 2016; 54: 2047 - 2050

Esposito S., Colombo C., Tosco A., Montemitro E., Volpi S., Ruggiero L., Lelii M., Bisogno A., Pelucchi C., Principi N. Streptococcus pneumoniae oropharyngeal colonization in children and adolescents with cystic fibrosis *Journal of Cystic Fibrosis 2016; 15: 366 - 371*

Green HD., Bright-Thomas RJ., Mutton KJ., Guiver M., Jones AM.

Increased prevalence of Pneumocystis jirovecii colonisation in acute pulmonary exacerbations of cystic fibrosis *Journal of Infection 2016; 73: 1 - 7*

Hector A., Kirn T., Ralhan A., Graepler-Mainka U., Berenbrinker S., Riethmueller J., Hogardt M., Wagner M., Pfleger A., Autenrieth I., Kappler M., Griese M., Eber E., Martus P., Hartl D.

Microbial colonization and lung function in adolescents with cystic fibrosis

Journal of Cystic Fibrosis 2016; 15: 340 - 349

Hendricks MR., Bomberger JM.

Digging through the Obstruction: Insight into the Epithelial Cell Response to Respiratory Virus Infection in Patients with Cystic Fibrosis

Journal of Virology 2016; 90: 4258 - 4261

Higgins G., Torre CF., Tyrrell J., McNally P., Harvey BJ., Urbach V.

Lipoxin A(4) prevents tight junction disruption and delays the colonization of cystic fibrosis bronchial epithelial cells by Pseudomonas aeruginosa

American Journal of Physiology-Lung Cellular and Molecular Physiology 2016; 310: L1053 - L1061

Hogan D.A., Willger S.D., Dolben E.L., Hampton T.H.,

Stanton B., Morrison H.G., Sogin M.L., Czum J., Ashare A. Analysis of lung microbiota in bronchoalveolar lavage, protected brush and sputum samples from subjects with Mild-to-Moderate cystic fibrosis lung disease PLoS ONE 2016; 11: 3:e0149998 -

Horsley A., Jones A.M., Lord R.

Antibiotic treatment for Burkholderia cepacia complex in people with cystic fibrosis experiencing a pulmonary exacerbation Cochrane Database of Systematic Reviews 2016; 2016: 1:CD009529

Ishiguro T., Takayanagi N., Baba Y., Takaku Y., Kagiyama N., Sugita Y.

Pulmonary Nontuberculous Mycobacteriosis and Chronic Lower Respiratory Tract Infections in Patients with Allergic Bronchopulmonary Mycosis without Cystic Fibrosis Internal Medicine 2016; 55: 1067 - 1070

Jani M., Mathee K., Azad RK.

Identification of Novel Genomic Islands in Liverpool Epidemic Strain of Pseudomonas aeruginosa Using Segmentation and Clustering

Frontiers in Microbiology 2016; 7: ArNo: 1210

Kamath KS., Pascovici D., Penesvan A., Goel A., Venkatakrishnan V., Paulsen IT., Packer NH., Molloy MP.

Pseudomonas aeruginosa Cell Membrane Protein Expression from Phenotypically Diverse Cystic Fibrosis Isolates Demonstrates Host-Specific Adaptations Journal of Proteome Research 2016; 15: 2152 - 2163

King J., Brunel SF., Warris A. Aspergillus infections in cystic fibrosis Journal of Infection 2016; 72:

Kumar B., Cardona ST.

Synthetic Cystic Fibrosis Sputum Medium Regulates Flagellar Biosynthesis through the flhF Gene in Burkholderia cenocepacia Frontiers in Cellular and Infection Microbiology 2016; 6: ArNo: 65

Limoli DH., Yang J., Khansaheb MK., Helfman B., Peng L., Stecenko AA., Goldberg JB.

Staphylococcus aureus and Pseudomonas aeruginosa coinfection is associated with cystic fibrosis-related diabetes and poor clinical outcomes

European Journal of Clinical Microbiology & Infectious Diseases 2016; 35: 947 - 953

Locke L.W., Myerburg M.M., Weiner D.J., Markovetz M.R., Parker R.S., Muthukrishnan A., Weber L., Czachowski M.R., Lacy R.T., Pilewski J.M., Corcoran T.E. Pseudomonas infection and mucociliary and absorptive clearance in the cystic fibrosis lung European Respiratory Journal 2016; 47: 1392 - 1401

Lund-Palau H., Turnbull AR., Bush A., Bardin E., Cameron

L., Soren O., Wierre-Gore N., Alton EWFW., Bundy JG., Connett G., Faust SN., Filloux A., Freemont P., Jones A., Khoo V., Morales S., Murphy R., Pabary R., Simbo A., et al Pseudomonas aeruginosa infection in cystic fibrosis: pathophysiological mechanisms and therapeutic approaches Expert Review of Respiratory Medicine 2016; 10: 685 - 697

Macdonald L.C., O'Keefe S., Parnes M.-F., Macdonald H., Stretz L., Templer S.J., Wong E.L., Berger B.W. A Secreted Ankyrin-Repeat Protein from Clinical

Stenotrophomonas maltophilia Isolates Disrupts Actin Cytoskeletal Structure ACS Infectious Diseases 2016; 2: 62 - 70

Madan JC.

Neonatal Gastrointestinal and Respiratory Microbiome in Cystic Fibrosis: Potential Interactions and Implications for Systemic Health

Clinical Therapeutics 2016; 38: 740 - 746

Maliniak ML., Stecenko AA., McCarty NA. A longitudinal analysis of chronic MRSA and Pseudomonas aeruginosa co-infection in cystic fibrosis: A single-center study Journal of Cystic Fibrosis 2016; 15: 350 - 356

McClean S., Healy ME., Collins C., Carberry S., O'Shaughnessy L., Dennehy R., Adams A., Kennelly H., Corbett JM., Carty F., Cahill LA., Callaghan M., English K., Bernard PMB., Doyle S., Shinoy M.

Linocin and OmpWAre Involved in Attachment of the Cystic Fibrosis-Associated Pathogen Burkholderia cepacia Complex to Lung Epithelial Cells and Protect Mice against Infection Infection and Immunity 2016; 84: 1424 - 1437

Mika M., Korten I., Qi WH., Regamey N., Frey U., Casaulta C., Latzin P., Hilty M.

The nasal microbiota in infants with cystic fibrosis in the first year of life: a prospective cohort study Lancet Respiratory Medicine 2016; 4: 627 - 635

Minandri F., Imperi F., Frangipani E., Bonchi C., Visaggio

D., Facchini M., Pasquali P., Bragonzi A., Visca P. Role of Iron Uptake Systems in Pseudomonas aeruginosa Virulence and Airway Infection Infection and Immunity 2016; 84: 2324 - 2335

Mitchell SL., Kajon AE., Kaplan SL., Kim J., Cardenas AM. An unusual case of disseminated adenovirus infection in a cystic fibrosis, liver transplant patient Journal of Clinical Virology 2016; 81: 64 - 67

Muniz FDM., Redondo MM., Sanchez CP., Rodriguez JG. Chronic lung infection caused by Trichosporon mycotoxinivorans and tricosporin mucoides in an immunocompetent cystic fibrosis patient

Archivos de Bronconeumologia 2016; 52: 400

Nguyen AT., Jones JW., Camara M., Williams P., Kane MA., Oglesby-Sherrouse AG.

Cystic Fibrosis Isolates of Pseudomonas aeruginosa Retain Iron-Regulated Antimicrobial Activity against Staphylococcus aureus through the Action of Multiple Alkylquinolones Frontiers in Microbiology 2016; 7: ArNo: 1171

Nielsen S., Needham B., Leach ST., Day AS., Jaffe A., Thomas T., Ooi CY.

Disrupted progression of the intestinal microbiota with age in children with cystic fibrosis Scientific Reports 2016; 6: ArNo: 24857

Padoan R., Poli P., Colombrita D., Borghi E., Timpano S., Berlucchi M.

Acute Scedosporium Apiospermum endobronchial infection in cystic fibrosis

Pediatric Infectious Disease Journal 2016; 35: 701 - 702

Pakasticali N., Kaya G., Senel U., Kipritci O., Tamay Z., Guler N., Nazik H., Ongen B.

Prevalence, antibiotic and pulsed-field gel electrophoresis patterns of staphylococcus aureus small-colony variants in cystic fibrosis patients

Southeast Asian Journal of Tropical Medicine and Public Health 2016; 47: 475 - 484

Power RF., Linnane B., Martin R., Power N., Harnett P., Casserly B., O'Connell NH., Dunne CP.

The first reported case of Burkholderia contaminans in patients with cystic fibrosis in Ireland: from the Sargasso Sea to Irish Children

BMC Pulmonary Medicine 2016; 16: ArNo: 57

Preece CL., Perry A., Gray B., Kenna DT., Jones AL., Cummings SP., Robb A., Thomas MF., Brodlie M., O'Brien CJ., Bourke SJ., Perry JD.

A novel culture medium for isolation of rapidly-growing mycobacteria from the sputum of patients with cystic fibrosis Journal of Cystic Fibrosis 2016; 15: 186 - 191

Preece CL., Wichelhaus TA., Perry A., Jones AL., Cummings SP., Perry JD., Hogardt M.

Evaluation of Various Culture Media for Detection of Rapidly Growing Mycobacteria from Patients with Cystic Fibrosis Journal of Clinical Microbiology 2016; 54: 1797 - 1803

Quinn RA., Phelan VV., Whiteson KL., Garg N., Bailey BA., Lim YW., Conrad DJ., Dorrestein PC., Rohwer FL. Microbial, host and xenobiotic diversity in the cystic fibrosis sputum metabolome

ISME Journal 2016; 10: 1483 - 1498

Qvist T., Taylor-Robinson D., Waldmann E., Olesen HV., Hansen CR., Mathiesen IH., Hoiby N., Katzenstein TL., Smyth RL., Diggle PJ., Pressler T.

Comparing the harmful effects of nontuberculous mycobacteria and Gram negative bacteria on lung function in patients with cystic fibrosis

Journal of Cystic Fibrosis 2016; 15: 380 - 385

Ramsay KA., Stockwell RE., Bell SC., Kidd TJ.

Infection in cystic fibrosis: impact of the environment and climate

Expert Review of Respiratory Medicine 2016; 10: 505 - 519

Reen FJ., Flynn S., Woods DF., Dunphy N., Chroinin MN., Mullane D., Stick S., Adams C., O'Gara F.

Bile signalling promotes chronic respiratory infections and antibiotic tolerance

Scientific Reports 2016; 6: ArNo: 29768

Saunders RV., Modha DE., Claydon A., Gaillard EA.

Chronic Aspergillus fumigatus colonization of the pediatric cystic fibrosis airway is common and may be associated with a more rapid decline in lung function *Medical Mycology 2016; 54: 537 - 543*

Schogler A., Stokes AB., Casaulta C., Regamey N., Edwards MR., Johnston SL., Jung A., Moeller A., Geiser T., Alves MP.

Interferon response of the cystic fibrosis bronchial epithelium to major and minor group rhinovirus infection

Journal of Cystic Fibrosis 2016; 15: 332 - 339

Schögler A., Stokes A.B., Casaulta C., Regamey N., Edwards M.R., Johnston S.L., Jung A., Moeller A., Geiser T., Alves M.P.

Interferon response of the cystic fibrosis bronchial epithelium to major and minor group rhinovirus infection Journal of Cystic Fibrosis 2016; 15: 332 - 339

Scoffield J., Silo-Suh L.

Glycerol metabolism promotes biofilm formation by Pseudomonas aeruginosa Canadian Journal of Microbiology 2016; 62: 704 - 710

Sommer LM., Alanin MC., Marvig RL., Nielsen KG., Hoiby N., von Buchwald C., Molin S., Johansen HK.

Bacterial evolution in PCD and CF patients follows the same mutational steps

Scientific Reports 2016; 6: ArNo: 28732

Sommer R., Wagner S., Varrot A., Nycholat CM., Khaledi A., Haussler S., Paulson JC., Imberty A., Titz A.

The virulence factor LecB varies in clinical isolates: consequences for ligand binding and drug discovery *Chemical Science* 2016; 7: 4990 - 5001

Sousa SA., Morad M., Feliciano JR., Pita T., Nady S., El-Hennamy RE., Abdel-Rahman M., Cavaco J., Pereira L., Barreto C., Leitao JH.

The Burkholderia cenocepacia OmpA-like protein BCAL2958: identification, characterization, and detection of anti-BCAL2958 antibodies in serum from B. cepacia complex-infected Cystic Fibrosis patients

AMB Express 2016; 6: ArNo: 41

Spadaro F., Scoffone VC., Chiarelli LR., Fumagalli M., Buroni S., Riccardi G., Forneris F.

The Crystal Structure of Burkholderia cenocepacia DfsA Provides Insights into Substrate Recognition and Quorum Sensing Fatty Acid Biosynthesis *Biochemistry 2016; 55: 3241 - 3250*

Stevens DA., Moss RB., Hernandez C., Clemons KV., Martinez M.

Effect of Media Modified To Mimic Cystic Fibrosis Sputum on the Susceptibility of Aspergillus fumigatus, and the Frequency of Resistance at One Center

Antimicrobial Agents and Chemotherapy 2016; 60: 2180 - 2184

Tabazavareh ST., Seitz A., Jernigan P., Sehl C., Keitsch S., Lang S., Kahl BC., Edwards M., Grassme H., Gulbins E., Becker KA.

Lack of Sphingosine Causes Susceptibility to Pulmonary Staphylococcus Aureus Infections in Cystic Fibrosis Cellular Physiology and Biochemistry 2016; 38: 2094 - 2102

Tang YJ., Ii B., Dai JG., Dai JF., Wang XH., Si J., Ali ZS., Li TT., He NY.

Genotyping of Pseudomonas aeruginosa Type III Secretion System Using Magnetic Enrichment Multiplex Polymerase Chain Reaction and Chemiluminescence Journal of Biomedical Nanotechnology 2016; 12: 762 - 769

van Mansfeld R., de Been M., Paganelli F., Yang L., Bonten M., Willems R.

Within-Host Evolution of the Dutch High-Prevalent Pseudomonas aeruginosa Clone ST406 during Chronic Colonization of a Patient with Cystic Fibrosis *PLoS One 2016; 11: 6:e0158106*

van Mansfeld R., de Vrankrijker A., Brimicombe R., Heijerman H., van Berkhout FT., Spitoni C., Grave S., van der Ent C., Wolfs T., Willems R., Bonten M.

The Effect of Strict Segregation on Pseudomonas aeruginosa in Cystic Fibrosis Patients

PLoS One 2016; 11: 6:e0157189

Vasiljevic ZV., Novovic K., Kojic M., Minic P., Sovtic A., Djukic S., Jovcic B.

Burkholderia cepacia complex in Serbian patients with cystic fibrosis: prevalence and molecular epidemiology *European Journal of Clinical Microbiology & Infectious Diseases* 2016; 35: 1277 - 1284

Wilton M., Wong MJQ., Tang L., Liang XY., Moore R., Parkins MD., Lewenza S., Dong TG.

Chelation of Membrane-Bound Cations by Extracellular DNA Activates the Type VI Secretion System in Pseudomonas aeruginosa

Infection and Immunity 2016; 84: 2355 - 2361

Winstanley C., O'Brien S., Brockhurst MA.

Pseudomonas aeruginosa Evolutionary Adaptation and Diversification in Cystic Fibrosis Chronic Lung Infections *Trends in Microbiology 2016; 24: 327 - 337*

Wong YC., Abd El Ghany M., Naeem R., Lee KW., Tan YC., Pain A., Nathan S.

Candidate Essential Genes in Burkholderia cenocepacia J2315 Identified by Genome-Wide TraDIS *Frontiers in Microbiology 2016; 7: ArNo: 1288*

Wood ME., Sherrard LJ., Ramsay KA., Yerkovich ST., Reid DW., Kidd TJ., Bell SC.

Methicillin-resistant Staphylococcus aureus acquisition in healthcare workers with cystic fibrosis: a retrospective crosssectional study

BMC Pulmonary Medicine 2016; 16: ArNo: 78

Wood M.E., Sherrard L.J., Ramsay K.A., Yerkovich S.T., Reid D.W., Kidd T.J., Bell S.C.

Methicillin-resistant Staphylococcus aureus acquisition in healthcare workers with cystic fibrosis: A retrospective crosssectional study

BMC Pulmonary Medicine 2016; 16: RF1:78 -

Yang ZS., Ma LQ., Zhu K., Yan JY., Bian L., Zhang KQ., Zou CG.

Pseudomonas toxin pyocyanin triggers autophagy: Implications for pathoadaptive mutations *Autophagy 2016; 12: 1015 - 1028* Zemanick ET., Hoffman LR.

Cystic Fibrosis Microbiology and Host Response Pediatric Clinics of North America 2016; 63: 617 - +

Nutrition

Alshaikh B., Schall JI., Maqbool A., Mascarenhas M., Bennett MJ., Stallings VA.

Choline supplementation alters some amino acid concentrations with no change in homocysteine in children with cystic fibrosis and pancreatic insufficiency

Nutrition Research 2016; 36: 418 - 429

Hanssens L., Thiebaut I., Lefevre N., Malfroot A., Knoop C., Duchateau J., Casimir G.

The clinical benefits of long-term supplementation with omega-3 fatty acids in cystic fibrosis patients - A pilot study Prostaglandins Leukotrienes and Essential Fatty Acids 2016; 108:45 - 50

Hauschild DB., Barbosa E., Moreira EAM., Neto NL., Platt VB., Filho P., Wazlawik E., Moreno YMF.

Nutrition Status Parameters and Hydration Status by Bioelectrical Impedance Vector Analysis Were Associated With Lung Function Impairment in Children and Adolescents With Cystic Fibrosis

Nutrition in Clinical Practice 2016; 31: 378 - 386

Isa H.

Re: Growth assessment and risk factors of malnutrition in children with cystic fibrosis Saudi Medical Journal 2016; 37: 712 - 713

Li L., Somerset S.

Dietary intake and nutritional status of micronutrients in adults with cystic fibrosis in relation to current recommendations Clinical Nutrition 2016; 35: 775 - 782

Lusman S., Sullivan J.

Nutrition and Growth in Cystic Fibrosis Pediatric Clinics of North America 2016; 63: 661 - +

Oliver C., Watson H.

Omega-3 fatty acids for cystic fibrosis Cochrane Database of Systematic Reviews 2016; : 1:CD002201

Simon MISD., Forte GC., Pereira JD., Procianoy EDA., Drehmer M.

Validation of a Nutrition Screening Tool for Pediatric Patients with Cystic Fibrosis

Journal of the Academy of Nutrition and Dietetics 2016; 116: 813 - 818

Simoneau T., Sawicki GS., Milliren CE., Feldman HA., Gordon CM.

A randomized controlled trial of vitamin D replacement strategies in pediatric CF patients Journal of Cystic Fibrosis 2016; 15: 234 - 241

Stark LJ., Opipari-Arrigan L., Filigno SS., Simon SL., Leonard A., Mogayzel PJ., Rausch J., Zion C., Powers SW. Web-Based Intervention for Nutritional Management in Cystic Fibrosis: Development, Usability, and Pilot Trial Journal of Pediatric Psychology 2016; 41: 510 - 521

Turck D., Braegger CP., Colombo C., Declercq D., Morton A., Pancheva R., Robberecht E., Stern M., Strandvik B.,

Wolfe S., Schneider SM., Wilschanski M. ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis Clinical Nutrition 2016; 35: 557 - 577

Woestenenk JW., Broos N., Stellato RK., Arets HGM., van der Ent CK., Houwen RHJ.

Vitamin A intake and serum retinol levels in children and adolescents with cystic fibrosis Clinical Nutrition 2016; 35: 654 - 659

Physiotherapy

Button BM., Wilson C., Dentice R., Cox NS., Middleton A., Tannenbaum E., Bishop J., Cobb R., Burton K., Wood M., Moran F., Black R., Bowen S., Day R., Depiazzi J., Doiron K., Doumit M., Dwyer T., Elliot A., Fuller L., Hall K., et al Physiotherapy for cystic fibrosis in Australia and New Zealand: A clinical practice guideline Respirology 2016; 21: 656 - 667

Feiten T.S., Flores J.S., Farias B.L., Rovedder P.M.E., Camargo E.G., Dalcin P.T.R., Ziegler B.

Respiratory therapy: A problem among children and adolescents with cystic fibrosis [Fisioterapia respiratória: Um problema de crianças e adolescentes com fibrose cística] Jornal Brasileiro de Pneumologia 2016; 42: 29 - 34

Vilozni D., Lavie M., Ofek M., Sarouk I., Bat-El Bar-Aluma., Dagan A., Ashkenazi M., Segel MJ., Efrati O. Consequences of Expiratory Flow Limitation at Rest in Subjects with Cystic Fibrosis

Annals of the American Thoracic Society 2016; 13: 825 – 832

Psychosocial

Calik-Kutukcu E., Saglam M., Vardar-Yagli N., Cakmak A., Inal-Ince D., Bozdemir-Ozel C., Sonbahar-Ulu H., Arikan H., Yalcin E., Karakaya J.

Listening to motivational music while walking elicits more positive affective response in patients with cystic fibrosis Complementary Therapies in Clinical Practice 2016; 23: 52 -58

Campbell F., Biggs K., Aldiss SK., O'Neill PM., Clowes M., McDonagh J., While A., Gibson F.

Transition of care for adolescents from paediatric services to adult health services

Cochrane Database of Systematic Reviews 2016; : 4:CD009794

Cavanaugh K., Read L., Dreyfus J., Johnson M., McNamara J.

Association of poor sleep with behavior and quality of life in children and adolescents with cystic fibrosis Sleep and Biological Rhythms 2016; 14: 199 - 204

Frederick C.

Psychosocial Challenges/Transition to Adulthood Pediatric Clinics of North America 2016; 63: 735 - +

Hoo ZH., Curley R., Campbell MJ., Walters SJ., Hind D., Wildman M.I.

Accurate reporting of adherence to inhaled therapies in adults with cystic fibrosis: methods to calculate "normative adherence" Patient Preference and Adherence 2016; 10: 887 - 900

Knudsen KB., Pressler T., Mortensen LH., Jarden M., Skov M., Quittner AL., Katzenstein T., Boisen KA.

Associations between adherence, depressive symptoms and health-related quality of life in young adults with cystic fibrosis Springerplus 2016; 5: ArNo: 1216

Olveira C., Sole A., Giron RM., Quintana-Gallego E., Mondejar P., Baranda F., Alvarez A., Prados C., Rodriguez-Gonzalez J., Herrero-Labarga I., Quittner A., Olveira G. Depression and anxiety symptoms in Spanish adult patients with cystic fibrosis: associations with health-related quality of life General Hospital Psychiatry 2016; 40: 39 - 46

Ortega-Garcia JA., Perales JE., Carceles-Alvarez A., Sanchez-Sauco MF., Villalona S., Mondejar-Lopez P., Pastor-Vivero MD., Escolano PM., Jaimes-Vega DC., Sanchez-Solis M.

Long term follow-up of a tobacco prevention and cessation program in cystic fibrosis patients Adicciones 2016; 28: 99 - 107

Pakhale S., Baron J., Armstrong M., Tasca G., Gaudet E., Aaron SD., Cameron W., Balfour L.

Lost in translation? How adults living with Cystic Fibrosis understand treatment recommendations from their healthcare providers, and the impact on adherence to therapy *Patient Education and Counseling 2016; 99: 1319 - 1324*

Sole A., Perez I., Vazquez I., Pastor A., Escriva J., Sales G., Hervas D., Glanville AR., Quittner AL.

Patient-reported symptoms and functioning as indicators of mortality in advanced cystic fibrosis: A new tool for referral and selection for Lung transplantation

Journal of Heart and Lung Transplantation 2016; 35: 789 - 794 Solem CT., Vera-Llonch M., Liu SZ., Botteman M.,

Castiglione B.

Impact of pulmonary exacerbations and lung function on generic health-related quality of life in patients with cystic fibrosis *Health and Quality of Life Outcomes 2016; 14: ArNo: 63*

Pulmonology

Beubler E., Fischer R., Untersteiner G., Strohmaier W.

Influence of the Surfactant Tyloxapol on Mucociliary Clearance in Human Respiratory Cystic Fibrosis Cells *Pharmacology 2016; 98: 1 - 3*

Brecelj J., Zidar N., Jeruc J., Orel R.

Morphological and Functional Assessment of Oesophageal Mucosa Integrity in Children With Cystic Fibrosis Journal of Pediatric Gastroenterology and Nutrition 2016; 62: 757 - 764

Chillappagari S., Muller C., Mahavadi P., Guenther A., Nahrlich L., Rosenblum J., Rubin BK., Henke MO.

A small molecule neutrophil elastase inhibitor, KRP-109, inhibits cystic fibrosis mucin degradation Journal of Cystic Fibrosis 2016; 15: 325 - 331

Goetz DM., Singh S.

Respiratory System Disease Pediatric Clinics of North America 2016; 63: 637 - +

Hamilos DL.

Chronic Rhinosinusitis in Patients with Cystic Fibrosis Journal of Allergy and Clinical Immunology-in Practice 2016; 4: 605 - 612

Heltshe SL., Szczesniak RD.

Forced expiratory volume in 1 second variability in cystic fibrosis- has the clinical utility been lost in statistical translation?

Journal of Pediatrics 2016; 172: 228

Kane M., Gonska T., Jensen R., Avolio J., Klingel M.,

Stanojevic S., Ratjen F. Lung clearance index response in patients with CF with class III CFTR mutations

Thorax 2016; 71: 476 - 477

Mastrigt E., Reyes-Reyes A., Brand K., Bhattacharya N., Urbach HP., Stubbs AP., de Jongste JC., Pijnenburg MW. Exhaled breath profiling using broadband quantum cascade laser-based spectroscopy in healthy children and children with asthma and cystic fibrosis

Journal of Breath Research 2016; 10: 2:026003

Morgan WJ., Pasta DJ., Konstan MW.

Forced expiratory volume in 1 second variability in cystic fibrosis- has the clinical utility been lost in statistical translation? Reply

Journal of Pediatrics 2016; 172: 228 - 229

Moss A., Juarez-Colunga E., Nathoo F., Wagner B., Sagel S. A comparison of change point models with application to longitudinal lung function measurements in children with cystic fibrosis

Statistics in Medicine 2016; 35: 2058 - 2073

Olszowiec-Chlebna M., Koniarek-Maniecka A., Stelmach W., Smejda K., Jerzynska J., Majak P., Bialasi M., Stelmach I.

Predictors of deterioration of lung function in Polish children with cystic fibrosis

Archives of Medical Science 2016; 12: 402 - 407

Quinn RA., Lim YW., Make TD., Whiteson K., Furlan M.,

Conrad D., Rohwer F., Dorrestein P. Metabolomics of pulmonary exacerbations reveals the personalized nature of cystic fibrosis disease *PEERJ 2016; 4: ArNo: e2174*

Radtke T., Puhan MA., Hebestreit H., Kriemler S.

The 1-min sit-to-stand test A simple functional capacity test in cystic fibrosis?

Journal of Cystic Fibrosis 2016; 15: 223 - 226

Reicher JJ., Mohabir P., Rad E., Gayer G. Increased prevalence of tracheal diverticula in cystic fibrosis patients

British Journal of Radiology 2016; 89: 1060:20150694

Smith D., Sovova K., Dryahina K., Dousova T., Drevinek P., Spanel P.

Breath concentration of acetic acid vapour is elevated in patients with cystic fibrosis

Journal of Breath Research 2016; 10: 2:021002

Stigliani M., Manniello MD., Zegarra-Moran O., Galietta L., Minicucci L., Casciaro R., Garofalo E., Incarnato L., Aquino RP., Del Gaudio P., Russo P.

Rheological Properties of Cystic Fibrosis Bronchial Secretion and in Vitro Drug Permeation Study: The Effect of Sodium Bicarbonate

Journal of Aerosol Medicine and Pulmonary Drug Delivery 2016; 29: 337 - 345

Stockley JA., Stockley RA.

Pulmonary Physiology of Chronic Obstructive Pulmonary Disease, Cystic Fibrosis, and Alpha-1 Antitrypsin Deficiency Annals of the American Thoracic Society 2016; 13:

VanDevanter DR., Morris NJ., Konstan MW.

IV-treated pulmonary exacerbations in the prior year: An important independent risk factor for future pulmonary exacerbation in cystic fibrosis *Journal of Cystic Fibrosis 2016; 15: 372 - 379*

Wells JM., Farris RF., Gosdin TA., Dransfield MT., Wood ME., Bell SC., Rowe SM.

Pulmonary artery enlargement and cystic fibrosis pulmonary exacerbations: a cohort study

Lancet Respiratory Medicine 2016; 4: 636 – 645

Radiology

Gergin O., Kawai K., MacDougall RD., Robson CD., Moritz E., Cunningham M., Adil E.

Sinus Computed Tomography Imaging in Pediatric Cystic Fibrosis: Added Value?

Otolaryngology-Head and Neck Surgery 2016; 155: 160 - 165

Jonczyk-Potoczna K., Nowak JK., Madry E., Katulska K., Stezowska-Kubiak S., Moczko J., Lisowska A., Walkowiak J.

Smaller Width of the Pancreatic Duct During Secretin-Enhanced Magnetic Resonance Cholangiopancreatography in Pancreatic-Sufficient Cystic Fibrosis Patients *Pancreas 2016; 45: 1175 -1183*

Oltmanns U., Palmowski K., Wielputz M., Kahn N., Baroke E., Eberhardt R., Wege S., Wiebel M., Kreuter M., Herth FJF., Mall MA.

Optical coherence tomography detects structural abnormalities of the nasal mucosa in patients with cystic fibrosis Journal of Cystic Fibrosis 2016; 15: 216 - 222

Theilmann RJ., Darquenne C., Elliott AR., Bailey BA., Conrad DJ.

Characterizing Lung Disease in Cystic Fibrosis with Magnetic Resonance Imaging and Airway Physiology *PLoS One 2016; 11: 6:e0157177*

Screening

Barben J., Rueegg CS., Jurca M., Spalinger J., Kuehni CE. Measurement of fecal elastase improves performance of newborn screening for cystic fibrosis *Journal of Cystic Fibrosis 2016; 15: 313 - 317*

Bergougnoux A., Boureau-Wirth A., Rouzier C., Altieri JP., Verneau F., Larrieu L., Koenig M., Claustres M., Raynal C. A false positive newborn screening result due to a complex allele carrying two frequent CF-causing variants

Journal of Cystic Fibrosis 2016; 15: 309 - 312

Castellani C., Massie J., Sontag M., Southern KW.

Newborn screening for cystic fibrosis Lancet Respiratory Medicine 2016; 4: 653 - 661

Destouni A., Poulou M., Kakourou G., Vrettou C., Tzetis M., Traeger-Synodinos J., Kitsiou-Tzeli S.

Single-cell high resolution melting analysis: A novel, generic, pre-implantation genetic diagnosis (PGD) method applied to cystic fibrosis (HRMA CF-PGD)

Journal of Cystic Fibrosis 2016; 15: 163 - 170

Grosse SD., Thompson JD., Ding Y., Glass M. The Use of Economic Evaluation to Inform Newborn Screening Policy Decisions: The Washington State Experience *Milbank Quarterly 2016; 94: 366 - 391*

Jessup M., Douglas T., Priddis L., Branch-Smith C., Shields L.

Parental Experience of Information and Education Processes Following Diagnosis of Their Infant With Cystic Fibrosis Via Newborn Screening Journal of Pediatric Nursing-nursing Care of Children &

Families 2016; 31: E233 - E241

Levy H., Nugent M., Schneck K., Stachiw-Hietpas D., Laxova A., Lakser O., Rock M., Dahmer MK., Biller J., Nasr SZ., Baker M., McColley SA., Simpson P., Farrell PM. Refining the continuum of CFTR-associated disorders in the era of newborn screening

Clinical Genetics 2016; 89: 539 - 549

Lundman E., Gaup HJ., Bakkeheim E., Olafsdottir EJ., Rootwelt T., Storrosten OT., Pettersen RD.

Implementation of newborn screening for cystic fibrosis in Norway. Results from the first three years Journal of Cystic Fibrosis 2016; 15: 318 - 324

Mak DYF., Sykes J., Stephenson AL., Lands LC. The benefits of newborn screening for cystic fibrosis: The Canadian experience

Journal of Cystic Fibrosis 2016; 15: 302 - 308

Salvatore M., Floridia G., Amato A., Censi F., Carta C., de Stefano MC., Ferrari G., Tosto F., Capoluongo E., Caruso U., Castaldo G., Cirilli N., Corbetta C., Padoan R., Raia V., Taruscio D.

The Italian pilot external quality assessment program for cystic fibrosis sweat test

Clinical Biochemistry 2016; 49: 601 - 605

Seror V., Cao C., Roussey M., Giorgi R.

PAP assays in newborn screening for cystic fibrosis: a population- based cost-effectiveness study Journal of Medical Screening 2016; 23: 62 - 69

Sontag MK., Lee R., Wright D., Freedenberg D., Sagel SD. Improving the Sensitivity and Positive Predictive Value in a Cystic Fibrosis Newborn Screening Program Using a Repeat Immunoreactive Trypsinogen and Genetic Analysis Journal of Pediatrics 2016; 175: 150 - +

Strekalova YA.

Finding Motivation: Online Information Seeking Following Newborn Screening for Cystic Fibrosis *Qualitative Health Research 2016; 26: 1180 - 1190*

Winter T., Muller C., Schmidt S., Nauck M.

3 years of newborn screening for cystic fibrosis in Mecklenburg- Western Pomerania. A field report for a nationwide implementation *Monatsschrift Kinderheilkunde 2016; 164: 591 - 597*

Zhang ZM., Lindstrom MJ., Farrell PM., Lai HCJ.

Pubertal Height Growth and Adult Height in Cystic Fibrosis After Newborn Screening *Pediatrics* 2016; 137: 5:e20152907

Therapy

Arora K., Yarlagadda S., Zhang WQ., Moon C., Bouquet E., Srinivasan S., Li CY., Stokes DC., Naren AP.

Personalized medicine in cystic fibrosis: genistein supplementation as a treatment option for patients with a rare S1045Y-CFTR mutation

American Journal of Physiology-lung Cellular and Molecular Physiology 2016; 311: L364 - L374

Aslam A., Sinha I.P., Southern K.W.

Ataluren and similar compounds (specific therapies for premature termination codon class I mutations) for cystic fibrosis

Cochrane Database of Systematic Reviews 2016; 2016: CD012040 -

Bali V., Lazrak A., Guroji P., Matalon S., Bebok Z. Mechanistic Approaches to Improve Correction of the Most Common Disease-Causing Mutation in Cystic Fibrosis *PLoS One 2016; 11: 5:e0155882*

Birket SE., Chu KK., Houser GH., Liu LB., Fernandez CM., Solomon GM., Lin V., Shastry S., Mazur M., Sloane PA., Hanes J., Grizzle WE., Sorscher EJ., Tearney GJ., Rowe SM.

Combination therapy with cystic fibrosis transmembrane conductance regulator modulators augment the airway functional microanatomy

American Journal of Physiology-Lung Cellular and Molecular Physiology 2016; 310: L928 - L939

Brandt C., Thronicke A., Roehmel JF., Krannich A., Staab D., Schwarz C.

Impact of Long-Term Tiotropium Bromide Therapy on Annual Lung Function Decline in Adult Patients with Cystic Fibrosis *PLoS One 2016; 11: 6:e0158193*

Craparo E.F., Porsio B., Sardo C., Giammona G., Cavallaro G.

Pegylated Polyaspartamide-Polylactide-Based Nanoparticles Penetrating Cystic Fibrosis Artificial Mucus Biomacromolecules 2016; 17: 767 - 777

De Boeck K., Amaral MD.

Progress in therapies for cystic fibrosis Lancet Respiratory Medicine 2016; 4: 662 - 674

Deeks ED.

Lumacaftor/Ivacaftor: A Review in Cystic Fibrosis Drugs 2016; 76: 1191 - 1201

Devereux G., Steele S., Griffiths K., Devlin E., Fraser-Pitt

D., Cotton S., Norrie J., Chrystyn H., O'Neil D. An Open-Label Investigation of the Pharmacokinetics and Tolerability of Oral Cysteamine in Adults with Cystic Fibrosis *Clinical Drug Investigation 2016; 36: 605 - 612*

Devereux G., Steele S., Griffiths K., Devlin E., Fraser-Pitt D., Cotton S., Norrie J., Chrystyn H., O'Neil D.

An Open-Label Investigation of the Pharmacokinetics and Tolerability of Oral Cysteamine in Adults with Cystic Fibrosis. *Clinical Drug Investigation*; 36: 605-812

Edmondson C., Davies JC.

Current and future treatment options for cystic fibrosis lung disease: latest evidence and clinical implications *Therapeutic Advances in Chronic Disease 2016; 7: 170 - 183*

Elborn JS., Ramsey BW., Boyle MP., Konstan MW., Huang XH., Marigowda G., Waltz D., Wainwright CE.

Efficacy and safety of lumacaftor/ivacaftor combination therapy in patients with cystic fibrosis homozygous for Phe508del CFTR by pulmonary function subgroup: a pooled analysis *Lancet Respiratory Medicine 2016; 4: 617 - 626*

Farinha C.M., Matos P.

Repairing the basic defect in cystic fibrosis - One approach is not enough

FEBS Journal 2016; 283: 246 - 264

Fila L., Bartakova LV., Grandcourtova A., Marel M., Drnek R., Bilkova A., Macek M., Drevinek P.

Ivacaftor in cystic fibrosis adults: Czech experience with six years of follow-up

Biomedical Papers-Olomouc 2016; 160: 276 - 279

Gaggar A., Chen JL., Chmiel JF., Dorkin HL., Flume PA.,

Griffin R., Nichols D., Donaldson SH. Inhaled alpha(1)-proteinase inhibitor therapy in patients with cystic fibrosis

Journal of Cystic Fibrosis 2016; 15: 227 - 233

Gelzo M., Sica C., Elce A., Dello Russo A., Iacotucci P., Carnovale V., Raia V., Salvatore D., Corso G., Castaldo G.

Carnovale V., Kaia V., Salvatore D., Corso G., Castaldo G. Reduced absorption and enhanced synthesis of cholesterol in patients with cystic fibrosis: a preliminary study of plasma sterols

Clinical Chemistry and Laboratory Medicine 2016; 54: 1461 - 1466

Haggie PM., Phuan PW., Tan JA., Zlock L., Finkbeiner WE., Verkman AS.

Inhibitors of pendrin anion exchange identified in a small molecule screen increase airway surface liquid volume in cystic fibrosis

FASEB Journal 2016; 30: 2187 - 2197

Halfhide C., Evans H.J., Couriel J.

Inhaled bronchodilators for cystic fibrosis Cochrane Database of Systematic Reviews 2016; 2016: CD003428 -

Haussermann S., Winnips C., Edelman J., Kappeler D., Herpich C., Ehlich H., Zanker D., Kietzig C., Sommerer K.

August C., Emirch H., Zahker D., Kleizig C., Sommerer K. Lung Deposition of Alpha(1)-Proteinase Inhibitor (Human) (A(1)-PI[H]) Inhalation Solution Using Two Inhalation Modes of the I-neb Adaptive Aerosol Delivery (AAD) System in Healthy Subjects and Subjects with Cystic Fibrosis Journal of Aerosol Medicine and Pulmonary Drug Delivery 2016; 29: 242 - 250

Islan G.A., Tornello P.C., Abraham G.A., Duran N., Castro G.R.

Smart lipid nanoparticles containing levofloxacin and DNase for lung delivery. Design and characterization

Colloids and Surfaces B: Biointerfaces 2016; 143: 168 - 176

Lands LC., Stanojevic S.

Oral non-steroidal anti-inflammatory drug therapy for lung disease in cystic fibrosis

Cochrane Database of Systematic Reviews 2016; : 4:CD001505 Mall MA.

Unplugging Mucus in Cystic Fibrosis and Chronic Obstructive Pulmonary Disease

Annals of the American Thoracic Society 2016; 13:

McColley SA.

A safety evaluation of ivacaftor for the treatment of cystic fibrosis

Expert Opinion on Drug Safety 2016; 15: 709 - 715

McElvaney NG.

Alpha-1 Antitrypsin Therapy in Cystic Fibrosis and the Lung Disease Associated with Alpha-1 Antitrypsin Deficiency Annals of the American Thoracic Society 2016; 13:

Mooney K., Ryan C., Downey DG.

Pharmacists' perspectives on monitoring adherence to treatment in Cystic Fibrosis

International Journal of Clinical Pharmacy 2016; 38: 296 - 302

Safe M., Gifford AJ., Jaffe A., Ooi CY.

Resolution of Intestinal Histopathology Changes in Cystic Fibrosis after Treatment with Ivacaftor Annals of the American Thoracic Society 2016; 13: 297 - 298

Thornton J., Rangaraj S.

Anti-inflammatory drugs and analgesics for managing symptoms in people with cystic fibrosis-related arthritis *Cochrane Database of Systematic Reviews 2016; : 1:CD006838* **Tűmmler B.**

Treatment of Cystic Fibrosis with CFTR Modulators Pneumologie 2016; 70: 301 - 313

Turnbull AR., Davies JC.

New drug developments in the management of cystic fibrosis lung disease

Expert Opinion on Pharmacotherapy 2016; 17: 1103 - 1112

van der Woerd WL., Wichers CGK., Vestergaard AL., Andersen JP., Paulusma CC., Houwen RHJ., van de Graaf SFJ.

Rescue of defective ATP8B1 trafficking by CFTR correctors as a therapeutic strategy for familial intrahepatic cholestasis Journal of Hepatology 2016; 64: 1339 - 1347

Virant-Young D., Thomas J., Woiderski S., Powers M., III, Carlier J., III, McCarty J., III, Kupchick T., III, Larder A., III

Cystic fibrosis: A novel pharmacologic approach to cystic fibrosis transmembrane regulator modulation therapy *Journal of the American Osteopathic Association 2015; 115: 546 - 555*

Yang C., Chilvers M., Montgomery M., Nolan SJ. Dornase alfa for cystic fibrosis

Cochrane Database of Systematic Reviews 2016; : 4:CD001127

Zhang W., Zhang X., Zhang YH., Stokes DC., Naren AP. Lumacaftor/ivacaftor combination for cystic fibrosis patients homozygous for phe508del-CFTR Drugs of Today 2016; 52: 229 – 237

Transplantation

Black S.M., Woodley F.W., Tumin D., Mumtaz K., Whitson B.A., Tobias J.D., Hayes D., Jr.

Cystic Fibrosis Associated with Worse Survival After Liver Transplantation

Digestive Diseases and Sciences 2016; 61: 1178 - 1185

Fakhro M., Ingemansson R., Skog I., Algotsson L., Hansson L., Koul B., Gustafsson R., Wierup P., Lindstedt S. 25-year follow-up after lung transplantation at Lund University

Hospital in Sweden: superior results obtained for patients with cystic fibrosis

Interactive Cardiovascular and Thoracic Surgery 2016; 23: 65 - 73

Faro A., Weymann A.

Transplantation

Pediatric Clinics of North America 2016; 63: 709 - +

Gautier SV., Golovinsky SV., Poptsov VN., Tsiroulnikova OM., Krasovsky SA., Mitrokhin AA., Vodneva MM., Sarygin PV., Akhaladze DG., Spirina EA.

Lung transplantation in cystic fibrosis patient with chronic airways infection of burkholderia cepacia (the first case in Russian Federation)

Vestnik Transplantologii I Iskusstvennyh Organov 2016; 18: 110 - 116

Hayes D., Kopp BT., Kirkby SE., Reynolds SD., Mansour HM., Tobias JD., Tumin D.

Impact of Donor Arterial Partial Pressure of Oxygen on Outcomes after Lung Transplantation in Adult Cystic Fibrosis Recipients

Lung 2016; 194: 547 - 553

Hayes D., Tumin D., Tobias JD.

Pre-transplant Panel Reactive Antibody and Survival in Adult Cystic Fibrosis Patients after Lung Transplantation Lung 2016; 194: 429 - 435

Karahasanovic A., Thorsteinsson AL., Bjarnason NH., Eiken P.

Long-term leukopenia in a lung transplanted patient with cystic fibrosis treated with zoledronic acid: a case report *Osteoporosis International 2016; 27: 2621 - 2625*

Moreno P., Alvarez A., Carrasco G., Redel J., Guaman HD.,

Baamonde C., Algar FJ., Cerezo F., Salvatierra A. Lung transplantation for cystic fibrosis: differential characteristics and outcomes between children and adults *European Journal of Cardio-Thoracic Surgery 2016; 49: 1334 -1343*

Pritchard J., Thakrar MV., Somayaji R., Surette MG., Rabin HR., Helmersen D., Lien D., Purighalla S., Waddell B., Parkins MD.

Epidemic Pseudomonas aeruginosa infection in patients with cystic fibrosis is not a risk factor for poor clinical Outcomes following lung transplantation

Journal of Cystic Fibrosis 2016; 15: 392 - 399

Ramos KJ., Quon BS., Psoter KJ., Lease ED., Mayer-Hamblett N., Aitken ML., Goss CH.

Predictors of non-referral of patients with cystic fibrosis for lung transplant evaluation in the United States

Journal of Cystic Fibrosis 2016; 15: 196 - 203

Schmid FA., Benden C.

Special considerations for the use of lung transplantation in pediatrics

Expert Review of Respiratory Medicine 2016; 10: 655 - 662 Special considerations for the use of lung transplantation in pediatrics

Expert Review of Respiratory Medicine 2016; 10: 655 - 662

Sivam S., Al-Hindawi Y., Di Michiel J., Moriarty C., Spratt P., Jansz P., Malouf M., Plit M., Pleass H., Havryk A., Bowen D., Haber P., Glanville AR., Bye PTP.

Liver and lung transplantation in cystic fibrosis: an adult cystic fibrosis centre's experience

Internal Medicine Journal 2016; 46: 852 - 854

Usatin DJ., Perito ER., Posselt AM., Rosenthal P.

Under Utilization of Pancreas Transplants in Cystic Fibrosis Recipients in the United Network Organ Sharing (UNOS) Data 1987-2014

American Journal of Transplantation 2016; 16: 1620 - 1625