**ECFS NEWSLETTER - Issue 58 - September 2018**

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**01. Letter from the President**

Dear Friends,

I hope this finds you well and that you all had a chance to enjoy some relaxing summer vacation. For those of you who participated in the June ECFS Conference in Belgrade, I am sure you will agree that it was a great success, and I would again like to thank the conference President Predrag Minic for his contribution to the event. I would also like to thank the local Cystic Fibrosis community and all the volunteers for 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 grants to young researchers based on the merit of their submitted abstracts, and we were happy to support 18 young researchers with a Travel Grant. We also granted 3 Young Investigator Awards and were delighted to welcome the Young Investigator Awardees to the Closing Ceremony. All the award winners also received a one-year free membership subscription to the ECFS.

**Young Investigator Awards 2018:**
On the left, Samuel Lara Reyna (UK) and Iris Silva (PT), on the right Mirjam Stahl (DE).
To honour the enormous contribution of Prof. Gerd Döring, the ECFS 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 Tavs Qvist (DK) for his manuscript about an antibody assay for diagnosing Mycobacterium abscessus published in Science in 2016.

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 on the homepage of our website www.ecfs.eu.

Kris De Boeck finished her mandate as ECFS president in June. Significant successes have been achieved for the Society under her leadership and we warmly thank her for her enthusiasm, dedication and vision.

Thanks go to all of you who participated to the Board elections. We are happy to announce that Jane Davies, Pavel Drevinek and Barry Plant have been elected as ECFS Board members. We also thank Daniel Peckham who finished his mandate as Board member and also congratulate him as he accepted to lead the ECFS Education Project. We also warmly thank Harm Tiddens for his commitment through the years as he finished his mandate as Secretary of the ECFS.

We would also like to warmly thank Tim Lee who has been a wonderful CTN Director and Pavel Drevinek who successfully led the Standards of Care project. We welcome Silke van Koningsbruggen-Rietschel and Carlo Castellani as the new Directors of CTN and the Standards of Care project.

We are well into the planning for the Basic Science Conference which will be held in Dubrovnik, Croatia 27-30 March 2019, and the Annual conference which will be held 05-08 June 2019 in Liverpool. 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 Liverpool will be 18 January 2019. 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 Belgrade, and you will find the minutes from the meeting included in this Newsletter.

Finally, I would like to draw your attention to the new General Data Protection Regulation (GDPR). The Regulation aims to reduce the collection of data from consumers without their knowledge and without transparency. We therefore need your consent to continue communicating with you. Please log in to your ECFS account, check your personal information profile settings so that we can continue sending you newsletters and information of interest to members.

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,

Isabelle Fajac, ECFS President
02. Annual General Meeting Minutes

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

41st European Cystic Fibrosis Conference, Belgrade, Serbia, 8th June 2018

Presiding Officer: Prof. Isabelle Fajac

The meeting was opened at 18.30.
Number of Participants: 57

President’s Report

Prof. Isabelle Fajac welcomed the ECFS members present and thanked them for their participation in the meeting. She thanked Prof. Kris De Boeck for her leadership of the ECFS in the last 3 years and commended her for being a person with a vision so inspirational to others.

She presented the agenda of the AGM and noted that reports on the current ECFS Projects and Working Groups 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 current figures of 1832 delegates registered for the Belgrade conference from 56 countries and 477 abstracts had been submitted. She underlined the importance of bringing the conference to this area of Europe and was happy at the numbers although lower than in previous years. She took the opportunity to thank the Conference President 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 this year, beginning with the Steering Committee meetings of the CTN, ECFSPR and Standards of Care in January and the ECFS Diagnostic Network Working Group meeting in St Gallen in February. She mentioned the highly successful Basic Science conference held in Loutraki, Greece where a record number of abstract submissions were received.

The President then presented the programme of activities for 2018-2019 to include:

- ECFS Board Autumn Meeting, Denver 17 October 2018
- ECFS Winter Board Meeting, Brussels 23 January 2019
- CTN Steering Group Meeting, Brussels 24-25 January 2019
- ECFS Standards of Care Meeting, Brussels 24 January 2019
- Registry Steering Group Meeting, Brussels 25 January 2019
- ECFS Diagnostic Network WG Meeting, Tunis 14-16 February 2019
- 16th ECFS Basic Science Conference, Dubrovnik 27-30 March 2019
- Young Investigators Meeting, Paris 27 Feb./01 March 2019
- 42nd European CF Conference, Liverpool 5-8 June 2019

Prof. Fajac presented the breadth of ECFS activities, including the major projects of the ECFS, the Working Groups and Special Interest Groups.

She highlighted recently approved Working Groups, the ECFS Fungal Pathogens Working Group and the ECFS Pulmonary Exacerbations Working Group and encouraged all interested to get in contact with the coordinators. She also mentioned continuing CFTR gene sequencing service with a goal to see 95% of patients with CF in every country to have 2 mutations identified.
Regarding the strategic plan to maximize progress in CF therapies, she announced that a document was made available for public comment and invited all to check the ECFS website and contribute by the deadline of 31 July 2018.

She was happy to report that the ECFS Book dedicated to infants and young children, The Early CF Years, was now published. She warmly thanked the editors, Kevin Southern and Kris De Boeck and all the authors of the different chapters. All ECFS members will receive a copy of the ECFS book by post.

Prof. Fajac presented some activities where active participation of ECFS members is strongly encouraged.

- Board elections
- ECFS Community bulletin boards (including job offers, collaborations asked and other announcements)
- Proposal box for ECFS members at the conference where suggestions for project opportunities and questions to ECFS board can be asked.
- Suggestions for tomorrow lounge activities
- Special interest groups associated with ECFS
- Social media

The audience was asked for any questions: none received

Secretary’s Report

Prof. Harm Tiddens reported the preliminary membership numbers for 2018 (until May) were 796, a substantial decrease in comparison with the figures reported at the same time frame in 2017 (986). One of the reasons for this could be the lower level of attendance to the conference and correlated lower number of members taking membership while registering. He encouraged all to advocate ECFS membership.

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 2019; 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 € (*)
- Retired members still actively engaged in an ECFS Project or Working Group - Free (*)
- Corporate membership rate for colleagues from the industry - 220 € (full membership benefits but no voting rights).

(*) 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 2019-2020-2021

The audience was asked for any questions: none received

Treasurer’s Report

Prof. Harm Tiddens presented the audited ECFS Financial statement for the year 2017. The result of the year was very favourable thanks to very good return on interests, with a surplus of € 201,423 resulting in a positive balance for the Society’s net assets of € 3,107,757 at 31st December 2017. He felt that keeping 2 to 3 years reserve was common practice and deemed that the level of expenses and commitment the Society has justified such a level of funds set aside. Prof. Tiddens informed the AGM that the full audited accounts will be posted on the website.

The audience was asked for any questions: none received

ECFS Board Elections

Prof. Fajac communicated the results of the recent elections. She thanked the departing Board members, Kris De Boeck, Harm Tiddens, Daniel Peckham for their commitment and support. She also thanked Pavel Drevinek for his leadership of the Standards of Care project. She was happy to announce the election of Prof. Jane Davies, Prof. Pavel Drevinek and Prof. Barry Plant as Board members. She also announced that the Board had decided to include Education as a major project of the ECFS and appointed Prof. Daniel Peckham as first Director for 3 years. Finally,
she was happy to announce that Dr. Carlo Castellani had agreed to take on the role of Director for the Standards of Care project.

*Update on Journal of Cystic Fibrosis*

Prof. Scott Bell reported on developments of JCF. There were 452 submissions and 152 Original Research Articles and Reviews published in 2017. There has been a steady increase in the number of Science Direct downloads (over 650,000).

New initiatives and recent changes include:

- Editorial board: retiring members are Isabelle Fajac and Martin Schwartz. Newly appointed as editors: Laura Sherrard, Pascale Fanen and Pierre-Regis Burgel
- Lancet Respiratory Medicine / JCF partnership: papers found more suitable to JCF than LRM sent to JCF
- Joint LRM/JCF symposium at conference: successful in 2016, 2017 and Belgrade 2018
- CF Research News: Very engaged with a total of 294 articles submitted and over 90% of JCF articles translated in lay language.

He showed a slide with the Top Cited articles from the start of JCF demonstrating the importance of guidelines to be published in JCF.

Finally, Prof. Bell underlined the need for enthusiastic reviewers for JCF.

The audience was asked for any questions:

- Lena Hjelte wondered whether the page limit should be set higher to allow more being published.
  Scott Bell specified that time between acceptance and print has been reduced from 10 months to 2 issues, which indicates that the page number is right.
- Vincent Gulmans announced that the CF Research News would be made more preeminent on CF Europe website and that the articles would be translated in more than 20 languages.

All felt this was important news and a very welcome initiative.

*Report from ECFS CTN*

Dr. Silke van Koningsbruggen-Rietschel thanked the ECFS CTN team and Executive committee for their commitment and hard work in a pleasant atmosphere. She also thanked all those committed in the different CTN committees for their hard work and dedication to the success of the CTN.

She presented a graph demonstrating the progression of studies conducted by the CTN since 2009. She also presented the evolution of the CTN services including number of protocols reviewed and number of feasibility checks performed. She reported 16 Protocol Reviews performed and 5 ongoing as of June 2018 and 5 Feasibilities finalized in May 2018 as well as 2 ongoing.

All CTN sites have received detailed weighted quality reports and metrics and low performing sites have been offered the possibility to enter a coaching programme before reapplying to be part of the CTN.

She thanked the Cystic Fibrosis Foundation for their support of the Additional Research Capacity Award and for funding quality management. She also thanked all CTN sites for data entry into the Trial Management System. She announced the new ECFS CTN expansion call for applications with a deadline of October 15 2018. Application form is available at ecfs-ctn@uzleuven.be.

The audience was asked for any questions: none received

*Report from ECFS Standards of Care*

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

The best practice subgroup reviewed and updated the 2014 Standards of Care. The document, ECFS Best practice guidelines: the 2018 revision, has now been published in open access. A report on a survey of Standards of care in Eastern Europe was also published recently.

The Quality Management subgroup finalised a quality management module for the Education platform. Also, a module for benchmarking across countries/centres is now released in the ECFSTracker.

The Standards of Care next steps will be:
- Quality Standards in adults to assess implementation of Standards of Care
- Benchmarking: dissemination
Pavel Drevinek is finishing his term in office and Carlo Castellani will be taking over from the AGM 2018. Finally, Pavel Drevinek thanked all the support during his term as Director of the Standards of Care and thanked Dr Carlo Castellani for taking on the leadership of this important project.

The audience was asked for any questions: none received

**Report from ECFS Patient Registry**

Dr. Luz Naehrlich reported on the activities of the ECFS Patient Registry (ECFSPR). He briefly presented the ECFSPR business plan for the years 2018-2020.

- Publish the ECFSPR Annual Report within 18 months of the close of a calendar year: He was happy to announce that the 2016 data report was published and reports 44719 patients in 31 countries.
- Strengthen the quality of data both in the ECFSPR database and at the site: An agreement is in place with the University of Mainz related to the development of Standard Operating Procedures and onsite monitoring visits.
- Increase scientific output: he announced a renewed and expanded Scientific Committee and several publications in the pipeline.
- Improve the software platform ECFSTracker: The platform will be updated to version 2.0 and available later in the year with the first hands on training sessions already taking place in Belgrade.
- Continue the cooperation with CF patient organisations: At-a-glance report and social media.
- Develop a standard operating procedure to handle and perform pharmacovigilance requests.

Finally, Dr Naehrlich thanked all partners and sponsors of the ECFSPR activities.

The audience was asked for any questions: none received

**Report from ECFS Education**

Prof. Daniel Peckham reported on the ECFS Education project. The aim of the ECFS Education is to strengthen the ECFS educational activities and streamline Education on Cystic Fibrosis.

An ECFS Education committee has been formed and includes 1 representative of each ECFS Working group, Special Interest Group and project (standards of care, CTN, Registry). The aims of the committee are to work closely with members and support the development of a clear curriculum appropriate to each sub speciality. The committee will advise on existing educational resources to be referenced, assess unmet educational needs and support the development of educational modules.

Terms of Reference for the group as well as standard procedures documents are being developed. Prof. Peckham also reported that the Education platform is set up and content continues being developed. At the Belgrade conference, the adult CF course, Nutrition masterclass, introductory course in Cognitive Behavioural Therapy will be filmed as well as a substantial number of symposia.

The audience was asked for any questions: none received

**Reports from ECFS Working Groups**

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

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
International Working Group on Antimicrobial Resistance
Fungal Pathogens Working Group
Pulmonary Exacerbations

The audience was asked for any questions: None received.

Reports from ECFS Special Interest Groups
Prior to the Annual General Meeting, members had been sent the progress reports from the ECFS Special Interest Groups.
ECFS Nurse Specialist Interest Group (ECFS NSiG)
European Cystic Fibrosis Pharmacy Group (ECFPG)
ECFS Psychosocial Working Group
European Cystic Fibrosis Nutrition Group

The audience was asked for any questions: None received.

Annual Conference 2019
A slide was shown presenting the 42nd ECFS conference, to be held in Liverpool, United Kingdom, 5-8 June 2019. Jane Davies, Conference President, invited all to attend the conference.

Any Other Business
None

With no further items being raised, the meeting ended at 19.30.
03. ECFS Board elections

The ECFS is happy to announce the results of the 2018 board elections. We had 3 positions open. Jane Davies, Pavel Drevinek and Barry Plant join the Board for a 3-year mandate.

04. ECFS Award - Dr. Susan Madge

In Belgrade, we were happy to hand over the ECFS Award to Dr. Susan Madge. This award acknowledges her remarkable contribution in the development of multidisciplinary care for people with cystic fibrosis. Su has lived through the transition of CF from a disease of childhood to predominantly a disease of adults. Her vision has been to ensure that the best care possible is available to people with CF of any age.

Once again, we extend our congratulations!

05. CTN - 3rd wave of expansion

The ECFS is pleased to announce a third wave of expansion of the ECFS-Clinical Trials Network (ECFS-CTN). The ECFS-CTN is a network of 43 Specialist Cystic Fibrosis Centres from 15 countries in Europe and Israel who are committed to a coordinated strategic approach to CF clinical trials in Europe and worldwide. This is done by sharing expertise, reviewing clinical trial protocols, maintaining high quality within sites, standardising outcome measures, validating new and alternative endpoints, providing training to the site’s staff and involving and cooperating with all stakeholders.

The ECFS is inviting further applications from CF Centres to become Clinical Research Centres in this Network. An Evaluation Board will be appointed by the Board of the ECFS. The Evaluation Board will be responsible for the ranking of the received applications.

The deadline for submission of the application form is October 15th 2018. The results of the selection will be officially announced at the ECFS conference in Liverpool, June 2019 and the new centres will actually join the network in January 2020.

Please feel free to forward this information to other centres that might be interested.

More information
06. In memoriam Prof. Peter Durie

Peter Durie, one of the world’s outstanding pediatric pancreatologists who had a crucial role in understanding the pancreatic and gastrointestinal pathology in CF passed away last month. He was born in Kenya and after his family returned to the UK, Peter graduated from the University of Wales in 1966 with an Honours degree in Zoology. He emigrated to Canada and began a career as a teacher. Then he made an ingenious decision to choose Judy, an English kindergarten teacher to be his wife. Without doubt she has been a pillar of strength and support throughout his life.

Peter then made a switch and entered Medical School, the McMaster Program in Hamilton, Ontario which had a new style of medical education. Dr. Richard Hamilton who was Division Chief at the Hospital for Sick Children wrote that the system there produced a generation of free-thinking troublemakers like Peter Durie! Peter joined the Pediatric Residency program in 1974 and subsequently entered Pediatric Gastroenterology at the Hospital for Sick Children.

Peter Durie was a Professor in the Department of Pediatrics, Faculty of Medicine University of Toronto and Staff Gastroenterologist, the Division of Gastroenterology, Hepatology and Nutrition and Senior Scientist in the Research Institute, the Hospital for Sick Children and was the Director of Cystic Fibrosis Research. Peter’s achievements at working at the interface between basic pathophysiology, front-line genetic research and clinical science made him the archetypal Clinician Scientist. He was an independent investigator at The Hospital for Sick Children with the prime focus of his research being the exocrine pancreas. He was the prime mover behind the pathophysiological studies of the pancreas in CF. After the CFTR gene was discovered in 1989 at the Hospital for Sick Children, Dr Durie characterized and demonstrated the genetic differences between PI and PS patients and made outstanding contributions in all aspects of CF including genetics, genotype/phenotype, CF liver disease, infertility and diabetes.

He was an ever-present at CF meetings around the world and was comfortable chairing basic science, gastroenterology or clinical nutrition sessions.

Throughout his career, Peter Durie maintained an exceedingly high level of commitment to patient care. He was a superb clinician and over 30 Fellows from all over the world learned a huge amount from him. He brought the bench to the bedside quicker than most and was truly loved by his patients and their families. He received many awards including the Paul di Saint Agnese Distinguished Scientific Achievement award from the North American CF Foundation.

He was my mentor and continued to support my career long after completion of Fellowship in Toronto. He was fond of the ECFS and a regular presenter for many years. He was very supportive of the setting up of the Diagnostic Network of ECFS and attended the initial meeting at ECFS.

He had an unparalleled integrity in research and caused more stringent guidelines on Industry supported research. Peter was a world leader in exocrine pancreatic research, a real ambassador for CF, an innovative thinker, a renowned educator and maybe even above all this, a genuinely kind, modest, compassionate human being and a model family man.

After terrible suffering in recent years he passed away on August the 27th.

He will be sorely missed by his wife, children and grandchildren and all those who had the privilege to know and learn from him.

Michael Wilschanski
07. CF Research News

With the CF Research News we want to provide access to patients, parents, relatives, friends and caregivers of patients with CF to all scientific work published in the Journal of Cystic Fibrosis (JCF). For more than 3 years, we publish every week lay version summaries of work published in the JCF. CF Europe and Cystic Fibrosis Australia also publish our articles on their websites, and several patient organisations do translate some of the articles in other languages. Recently, a few colleagues from the US also provided us lay version summaries of papers they have published in other journals and we are happy to also post them on our page as it contributes to bridge the gap between people with CF and the researchers investigating CF.

08. 13th European CF Young Investigators Meeting

The ECFS and the Patients organisations will jointly organise the 13th European CF Young Investigators Meeting at the Institute Pasteur in Paris on 27 February - 01 March 2019. It aims at fostering interactions between young scientists in order to create a long-term collaborative European network of CF investigators. The meeting will cover all CF research fields, considering basic research and clinical research. 

More information

Deadline for application: 04 November 2018

09. Upcoming Events

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<th>Location</th>
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<td>Denver, CO, US</td>
<td>17 October 2018</td>
<td>ECFS Board Meeting</td>
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<td>18-20 October 2018</td>
<td>33rd North American Cystic Fibrosis Conference</td>
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<td>Brussels, Belgium</td>
<td>23 January 2019</td>
<td>ECFS Winter Board Meeting</td>
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<td>24 January 2019</td>
<td>ECFS Standards of Care Meeting</td>
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<td>24-25 January 2019</td>
<td>ECFS CTN Steering Group Meeting</td>
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<td>25 January 2019</td>
<td>ECFS Patient Registry Steering Group Meeting</td>
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<td>Tunis, Tunisia</td>
<td>14-16 February 2019</td>
<td>ECFS Diagnostic Network Working Group Meeting</td>
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<td>Paris, France</td>
<td>27-February - 01 March 2019</td>
<td>13th European Young Investigators Meeting</td>
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<td></td>
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<td>Abstract submission from 01 October to 04 November 2018</td>
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<td>Liverpool, UK</td>
<td>04 June 2019</td>
<td>ECFS Board Meeting</td>
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<td>05-08 June 2019</td>
<td>42nd European CF Conference</td>
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Dubrovnik, Croatia  
27-30 March 2019  
ECFS 16th Basic Science Conference
10. Deadlines

- Comments Lung Transplant Referral for Individuals with Cystic Fibrosis: CFF Consensus Guidelines, 10 October 2018
- Abstract submission Young Investigator Meeting 04 November 2018
- Abstract submission 16th ECFS Basic Science conference 11 January 2019
- Abstract submission ECFS Conference Liverpool 18 January 2019
- Nomination ECFS Award 15 February 2019
- Nomination Gerd Döring Award 15 February 2019
- Nomination ECFS Elections 30 March 2019

11. 2018 Belgrade Conference Survey - Free Registration Winners

The ECFS would like to thank all those who took the time to answer the ECFS 2018 Survey after the conference in Belgrade. The answers and comments help us to keep on improving our conferences.

As announced, three free registrations to the next ECFS Conference in Liverpool were selected from the respondents.

We congratulate the winners:

- Anita Senstad Wathne, Norway
- Snežana Živanović, Serbia
- Nicolas Richard, France

12. Vertex Innovation Awards (VIA)

The Vertex Innovation Awards (VIA) Committee is pleased to announce that applications are now open for the VIA 2019 awards programme to support innovative clinical research that may improve the care of patients with cystic fibrosis.

The VIA Committee will select up to four projects to support with grants up to a total of €750,000.

Deadline for submission: 09 November 2018

FIND OUT MORE: http://vertexinnovationawards.com/

13. Current references in CF

Please scroll to next page for full list.
CFTR

Ahmadi S., Xia S., Wu YS., Di Paola M., Kisson R., Luk C., Lin F., Du K., Rommens J., Bear CE. SLC6A14, an amino acid transporter, modifies the primary CF defect in fluid secretion ELife 2018; 7: ArtNo: e37963

Aleksejrov LA., Fay JF., Riordan JR. R- Domain Phosphorylation by Protein Kinase A Stimulates Dissociation of Unhydrolyzed ATP from the First Nucleotide-Binding Site of the Cystic Fibrosis Transmembrane Conductance Regulator Biochemistry 2018; 57: 5073 - 5075

Brewington JJ., Filbrandt ET., LaRosa FJ., Moncivaiz JD., Ostmann AJ., Strecker LM., Clancy JP. Brushed nasal epithelial cells are a surrogate for bronchial epithelial CFTR studies JCI Insight 2018; 3: 13:e69385

Bruch BA., Singh SB., Ramsey LJ., Starner TD. Impact of a cystic fibrosis transmembrane conductance regulator (CFTR) modulator on high-dose ibuprofen therapy in pediatric cystic fibrosis patients Pediatric Pulmonology 2018; 53: 1035 - 1039


Callebaut I., Chong PA., Forman JDG., Farinha CM., Miller E., McCarty N. Aims Genetics 2018; 5: 53

Capecci JM., Loguerio S., Roth DM., Su AL., Balch WE. Correcting the F508del-CFTR variant by modulating eukaryotic translation initiation factor 3-mediated translation initiation Journal of Biological Chemistry 2018; 293: 13477 - 13495

Hutt DM., Mishra SK., Roth DM., Larsen MB., Angles F., Frizzell RA., Balch WE. Silencing of the Hsp70-specific nucleotide-exchange factor BAG3 corrects the F508del-CFTR variant by restoring autophagy Journal of Biological Chemistry 2018; 293: 13082 - 13095

Kramer EL., Hardie WD., Madala SK., Davidson C., Clancy JP. Subacute TGF beta expression drives inflammation, goblet cell hyperplasia, and pulmonary function abnormalities in mice with effects dependent on CFTR function American Journal of Physiology-lung Cellular and Molecular Physiology 2018; 315: L455 - L465


Laselva O., Molinski S., Casavola V., Bear CE. Correc tors of the Major Cystic Fibrosis Mutant Interact through Membrane-Spanning Domains Molecular Pharmacology 2018; 93: 612 - 618

Linsdell P. Cystic fibrosis transmembrane conductance regulator (CFTR): Making an ion channel out of an active transporter structure Channels 2018; 12: 284 - 290

Malhotra KA., Noor MO., Krull UJ. Detection of cystic fibrosis transmembrane conductance regulator Delta F508 gene mutation using a paper-based nucleic acid hybridization assay and a smartphone camera Analyst 2018; 143: 3049 - 3058

Martin SL., Saint-Criq V., Hwang TC., Csamady L. Ion channels as targets to correct cystic fibrosis lung disease Journal of Cystic Fibrosis 2018; 17:

Matos AM., Gomes-Duarte A., Faria M., Barros P., Jordan P., Amaral MD., Matos P. Prolonged co-treatment with HGF sustains epithelial integrity and improves pharmacological rescue of Phe508del-CFTR Scientific Reports 2018; 8: ArtNo: 13026

Matthes E., Hanrahan JW., Cantin AM. F508del-CFTR is not corrected by thymosin alpha 1 Nature Medicine 2018; 24: 890 - 891


Mutolo MJ., Leir SH., Fossum SL., Browne JA., Harris A. A transcription factor network represses CFTR gene expression in airway epithelial cells Biochemical Journal 2018; 475: 1323 - 1334

Negoda A., Cowley EA., El Hiani Y., Linsdell P. Conformational change of the extracellular parts of the CFTR protein during channel gating Cellular and Molecular Life Sciences 2018; 75: 3027 - 3038


Numata T., Sato-Numata K., Okada Y., Inoue R. Cellular mechanism for herbal medicine Junchoto to facilitate intestinal Cl-/water secretion that involves cAMP-dependent activation of CFTR Journal of Natural Medicines 2018; 72: 694 - 705
Databases & Registries

Ahern S., Sims G., Earnest A., Bell SC.
Optimism, opportunities, outcomes: the Australian Cystic Fibrosis Data Registry
Internal Medicine Journal 2018; 48: 721 - 723

Data from the US and UK cystic fibrosis registries support disease modification by CFTR modulation with ivacaftor
Thorax 2018; 73: 731 - 740

Dansenbrook EC., Sawicki GS.
Cystic fibrosis patient registries: A valuable source for clinical research
Journal of Cystic Fibrosis 2018; 17: 433 - 440

Hoo ZH., Curley R., Campbell MJ., Walters SJ., Wildman MJ.
The importance of data issues when comparing cystic fibrosis registry outcomes between countries: Are annual review FEV1 in the UK only collected when subjects are well?
Journal of Evaluation in Clinical Practice 2018; 24: 745 - 751

Hurley PD., Oliver S., Mehta A.
Creating longitudinal datasets and cleaning existing data identifiers in a cystic fibrosis registry using a novel Bayesian probabilistic approach from astronomy

Diabetes

Armaghanian N., Markovic TP., Brand-Miller JC., Bye PTP., Moriarty CP., Steinbeck KS.
Hypoglycaemia in cystic fibrosis: An analysis of a single centre adult cystic fibrosis clinic
Journal of Cystic Fibrosis 2018; 17: 542 - 547

Bridges N., Rowe R., Holt RIG.
Unique challenges of cystic fibrosis-related diabetes
Diabetic Medicine 2018; 45: 1181 - 1188

Chan CL., Hope E., Thurston J., Vigers T., Pyle L., Zeitler PS., Nadeau KJ.
Hemoglobin A(1c) Accurately Predicts Continuous Glucose Monitoring-Derived Average Glucose in Youth and Young Adults With Cystic Fibrosis
Diabetes Care 2018; 41: 1406 - 1413

Machura E., Szczepanska M., Swietochowska E., Halkiewicz F., Barc-Czarnecka M., Ziora K., Ziora D.
Evaluation of adipokines in children with cystic fibrosis
International Journal of Obesity 2018; 12: 585 - 593

Mettstein M.
Diego: a life with cystic fibrosis
Manuelle Therapie 2018; 22: 67 - 73

Whitehead L., Arabiat RND., Foster M.
Singing as an adjunct therapy for children and adults with cystic fibrosis: A Cochrane review summary
Archives of Disease in Childhood 2018; 103: 592 - 596

Glucose trajectories in cystic fibrosis and their association with pulmonary function
Journal of Cystic Fibrosis 2018; 17: 400 - 406
Diagnosis

Bergougnoux A., Taulan-Cadar M., Claustres M., Raynal C.
Current and future molecular approaches in the diagnosis of cystic fibrosis

Intra-individual biological variation in sweat chloride concentrations in CF, CFTR dysfunction, and healthy pediatric subjects
*Pediatric Pulmonology* 2018; 53: 728 - 734

Cirilli N., Southern KW., Buzzetti R., Barben J., Nahrlich L., Munck A., Wilschanski M., De Boeck K., Derichs N.
Real life practice of sweat testing in Europe
*Journal of Cystic Fibrosis* 2018; 17: 325 - 332

Vallejos S., Hernando E., Trigo M., Garcia FC., Garcia-Valverde M., Iurbe D., Cabero MJ., Quesada R., Garcia JM.
Polymeric chemosensor for the detection and quantification of chloride in human sweat. Application to the diagnosis of cystic fibrosis
*Journal of Materials Chemistry b* 2018; 6: 3735 - 3741

Epidemiology

Boussetta K., Khaibi F., Bahri Y., Belhadj L., Tinsa F., Ben Messaoud T., Hamouda S.
Cystic fibrosis in Tunisian children: a review of 32 children
*African Health Sciences* 2018; 18: 664 - 670

Grangeia A., Alves S., Goncalves L., Gregorio L, Santos AC., Barros H., Barros A., Carvalho F., Mora C.
Spectrum of CFTR gene sequence variants in a northern Portuguese population
*Pulmonology* 2018; 24: 3 - 9

Jackson AD., Goss CH.
Epidemiology of CF: How registries can be used to advance our understanding of the CF population
*Journal of Cystic Fibrosis* 2018; 17: 297 - 305

Jarjour RA., Al-Berrawi S., Ammar S., Majdalawi R.
Spectrum of cystic fibrosis mutations in Syrian patients
*Minerva Pediatrica* 2018; 70: 159 - 164

Exercise

Supervised physical exercise improves clinical, anthropometric and biochemical parameters in adult cystic fibrosis patients: A 2-year evaluation
*Clinical Respiratory Journal* 2018; 12: 2228 - 2234

Gruet M., Mely L., Vallier JM.
Overall and differentiated sensory responses to cardiopulmonary exercise test in patients with cystic fibrosis: kinesthetics and ability to predict peak oxygen uptake

Paranjape SM., Carson KA., Demissie SM., Loosin H., Vela K., Mogayzel PJ.
Use of the Modified Shuttle Walk Test During Inpatient Pediatric Cystic Fibrosis Pulmonary Exacerbation Treatment
*Journal of Acute Care Physical Therapy* 2018; 9: 136 - 142

Pfirrmann D., Haller N., Huber Y., Jung P., Lieb K., Gockel L., Poplawksa K., Schattenberg JM., Simon P.
Applicability of a Web-Based, Individualized Exercise Intervention in Patients With Liver Disease, Cystic Fibrosis, Esophageal Cancer, and Psychiatric Disorders: Process Evaluation of 4 Ongoing Clinical Trials
*JMIR Research Protocols* 2018; 7: 5:e106

Sovtic A., Minic P., Markovic-Sovtic G., Trajkovic GZ.
Respiratory Muscle Strength and Exercise Performance in Cystic Fibrosis-A Cross Sectional Study
*Frontiers in Pediatrics* 2018; 6: ArtNo: 244

Gastroenterology

Bolia R., Ooi CY., Lewindon P., Bishop J., Ranganathan S., Harrison J., Ford K., van der Haak N., Oliver MR.
Practical approach to the gastrointestinl manifestsations of cystic fibrosis
*Journal of Paediatrics and Child Health* 2018; 54: 609 - 619

Lack of efficacy of Lactobacillus GG in reducing pulmonary exacerbations and hospital admissions in children with cystic fibrosis: A randomised placebo controlled trial
*Journal of Cystic Fibrosis* 2018; 17: 375 - 382

Kent DS., Remer T., Blumenthal C., Hunt S., Simonds S., Egert S., Gaskin KJ.
C-13-Mixed Triglyceride Breath Test and Fecal Elastase as an Indirect Pancreatic Function Test in Cystic Fibrosis Infants
*Journal of Pediatric Gastroenterology and Nutrition* 2018; 66: 811 - 815

Maisonneuve P., Lowenfels AB., Hadjiliadis D., Khoruts A., Marshall BC.
Gastrointestinal cancers in patients with cystic fibrosis
*Lancet Oncology* 2018; 19: E368

Kent DS., Remer T., Blumenthal C., Hunt S., Simonds S., Egert S., Gaskin KJ.
C-13-Mixed Triglyceride Breath Test and Fecal Elastase as an Indirect Pancreatic Function Test in Cystic Fibrosis Infants
*Journal of Pediatric Gastroenterology and Nutrition* 2018; 66: 811 - 815

Ratchford TL., Teckman JH., Patel DR.
Gastrointestinal pathophysiology and nutrition in cystic fibrosis

Yamada A., Komaki Y., Komaki F., Micic D., Zullo S., Sakuraba A.
Risk of gastrointestinal cancers in patients with cystic fibrosis: a systematic review and meta-analysis
*Lancet Oncology* 2018; 19: 758 - 767

Yamada A., Komaki Y., Komaki F., Micic D., Zullo S., Sakuraba A.
Gastrointestinal cancers in patients with cystic fibrosis reply
*Lancet Oncology* 2018; 19: E369
Kaluzna-Czyz M., Grzybowska-Chlebowczyk U., Wos H., Wiecek S. 
Serum Heparin Level as a Marker of Iron Status in Children with Cystic Fibrosis

Mediators of Inflammation 2018; : ArtNo: 3040346

Neutrophil elastase correlates with increased sphingolipid content in cystic fibrosis sputum
Pediatric Pulmonology 2018; 53: 872 - 880

Kunnurarapuru AB., Afosah DK., Sankaranaranayan NV., Gangji RN., Zheng S., Kennedy T., Rubin BK., Voynow JA., Desai UR. 
Molecular principles for heparin oligosaccharide-based inhibition of neutrophil elastase in cystic fibrosis
Journal of Biological Chemistry 2018; 293: 12480 - 12490

Liu C., Pan XL., Xia B., Chen F., Jin YX., Bai F., Pieeie G., Cheng ZH., Jin SG., Wu WH. 
Construction of a Protective Vaccine Against Lipopolysaccharide-Heterologous Pseudomonas aeruginosa Strains Based on Expression Profiling of Outer Membrane Proteins During Infection
Frontiers in Immunology 2018; 9: ArtNo: 1737

Manji J., Thamboo A., Tacey M., Garnis C., Chadha NK. 
The presence of Interleukin-13 in nasal lavage may be a predictor of nasal polyposis in pediatric patients with cystic fibrosis
Rhinology 2018; 56: 261 - 267

Mauch RM., Norregaard LL., Ciofu O., Levy CE., Hoiby N. 
IgG avidity to Pseudomonas aeruginosa over the course of chronic lung biofilm infection in cystic fibrosis
Journal of Cystic Fibrosis 2018; 17: 356 - 359

Montuschi P., Lucidi V., Paris D., Montemiro E., Shohreh R., Mores N., Melck D., Santini G., Majo F., Motta A. 
Metabolomic Analysis by Nuclear Magnetic Resonance Spectroscopy as a New Approach to Understanding Inflammation and Monitoring of Pharmacological Therapy in Children and Young Adults With Cystic Fibrosis
Frontiers in Pharmacology 2018; 9: ArtNo: 595

Scalbert-Dujardin M., Boldron A., Leroy E., Bazin J., Froment-Leclercq E. 
Influenza vaccination and cystic fibrosis. Impact of an incentivisation campaign about influenza vaccination for patients attending the Dunkerque cystic fibrosis treatment centre and their health care workers
Revue des Maladies Respiratoires 2018; 35: 279 - 286

Sheehan G., Bergsson G., McElvaney NG., Reeves EP., Kavanagh K. 
The Human Cathelicidin Antiinflammatory Peptide LL-37 Promotes the Growth of the Pulmonary Pathogen Aspergillus fumigatus
Infection and Immunty 2018; 86: 7:e00997-18

Sousa SA., Seixas AMM., Leitao JH. 
Postgenomic Approaches and Bioinformatics Tools to Advance the Development of Vaccines against Bacteria of the Burkholderia cepacia Complex
Vaccines 2018; 6: 2:34

Opposite Expression of Hepatic and Pulmonary Corticosteroid-Binding Globulin in Cystic Fibrosis Patients
Frontiers in Pharmacology 2018; 9: ArtNo: 545

Wilton M., Halverson TWR., Charron-Mazenod L., Parkins MD., Lewenza S. 
Secreted Phosphatase and Deoxyribonuclease Are Required by Pseudomonas aeruginosa To Defend against Neutrophil Extracellular Traps
Infection and Immunty 2018; 86: 9:e00403-18

Meeting Abstracts or Reports

Amaral MD., Boj SF., Shaw J., Leiziger J., Beckman JM. 
Cystic fibrosis: Beyond the airways. Report on the meeting of the basic science working group in Loutraki, Greece
Journal of Cystic Fibrosis 2018; 17: 441 - 443

Mall MA., Hwang TC., Braaikman I. 
Cystic fibrosis research topics featured at the 14th ECFS Basic Science Conference: Chairman's summary
Journal of Cystic Fibrosis 2018; 17:

Martiniano SL., Toprak D., Ong T., Zemanick ET., Daines CL., Muhlebach MS., Esther CR., Dellen EP. 
Highlights from the 2017 North American Cystic Fibrosis Conference
Pediatric Pulmonology 2018; 53: 979 - 986

Microbiology

Airway persistence by the emerging multi-azole-resistant Rasamsonia argillacea complex in cystic fibrosis
Mycoses 2018; 61: 665 - 673

Adjemian J., Olivier KN., Prevots DB. 
Epidemiology of Pulmonary Nontuberculous Mycobacterial Sputum Positivity in Patients with Cystic Fibrosis in the United States, 2010-2014

Ahmed MN., Porse A., Sommer MOA., Hoiby N., Ciofu O. 
Evolution of Antibiotic Resistance in Biofilm and Planktonic Pseudomonas aeruginosa Populations Exposed to Subinhibitory Levels of Ciprofloxacin
Antimicrobial Agents and Chemotherapy 2018; 62: 8:e00320-18

Aiello TB., Levy CE., Zaccariotto RT., Paschold IA., Pereira MC., da Silva MTN., Ribeiro JD., Ribeiro AF., Toro AACD., Mauch RM. 
Prevalence and clinical outcomes of nontuberculous mycobacteria in a Brazilian cystic fibrosis reference center
Pathogens and Disease 2018; 76: 5:fty051

Alhamzi A. 
NOD-like receptor(s) and host immune responses with Pseudomonas aeruginosa infection
Inflammation Research 2018; 67: 479 - 493

Bardin EE., Cameron SJS., Perdones-Montero A., Hardiman K., Bolt F., Alton EFWF., Bush A., Davies JC., Takats Z. 
Metabolic Phenotyping and Strain Characterisation of Pseudomonas aeruginosa isolates from Cystic Fibrosis Patients Using Rapid Evaporative Ionisation Mass Spectrometry
Scientific Reports 2018; 8: ArtNo: 10952

Barlow G., Morice A. 
Successful treatment of resistant Burkholderia multivorans infection in a patient with cystic fibrosis using ceftazidime/avibactam plus aztreonam
Journal of Antimicrobial Chemotherapy 2018; 73: 2270 - 2271

Borisova D., Strateva T., Paunova-Kristeva T., Mitov L., Stoilova S. 
Phenotypic investigation of paired pseudomonas aeruginosa strains isolated from cystic fibrosis patients prior- and post-tobramycin treatment
Comptes Rendus de L Academie Bulgare des Sciences 2018; 71: 1044 - 1051

Boutin S., Weitzmann M., Hassel S., Graeber SY., Stahl M., Dittrich AS., Mall MA., Dalpeke AH. 
One time quantitative PCR detection of Pseudomonas aeruginosa to discriminate intermittent from chronic infection in cystic fibrosis
Journal of Cystic Fibrosis 2018; 17: 348 - 355
Hahn A., Bendall ML., Gibson KM., Chaney H., Sami I., Perez GF., Koumbourlis AC., McCaffrey TA., Freistat RJ., Crandall KA.

Benchmark Evaluation of True Single Molecular Sequencing to Determine Cystic Fibrosis Airway Microbiome Diversity Frontiers in Microbiology 2018; 9: 1069

Heacock-Kang Y., Zarzyczy-Siek J., Sun Z., Poonsuk K., Bluhm AP., Cabanas D., Fogen D., McMillan IA., Chuanchuen R., Hoang TT.

Novel dual regulators of Pseudomonas aeruginosa essential for productive biofilms and virulence Molecular Microbiology 2018; 109: 401 - 414

Heltshen SL., Khan U., Beckett V., Baines A., Emerson J., Sanders DB., Gibson RL., Morgan W., Rosenfeld M.

Longitudinal development of initial, chronic and mucoid Pseudomonas aeruginosa infection in young children with cystic fibrosis Journal of Cystic Fibrosis 2018; 17: 341 - 347

Klockgether J., Cramer N., Fischer S., Wiehlmann L., Tummeler B.

Long-Term Microevolution of Pseudomonas aeruginosa Differs between Mildly and Severely Affected Cystic Fibrosis Lungs American Journal of Respiratory Cell and Molecular Biology 2018; 59: 246 - 256

Lore NL., Cigan C., Sipione B., Dragonzi A.

The impact of host genetic background in the Pseudomonas aeruginosa respiratory infections Mammalian Genome 2018; 29: 550 - 557

Lucca F., Guarnieri M., Mos R., Muffato G., Rigoli R., Da Dalt L.

Antibiotic resistance evolution of Pseudomonas aeruginosa in cystic fibrosis patients (2010-2013) Clinical Respiratory Journal 2018; 12: 2189 - 2196

Mahomed TG., Kock MM., Masekela R., Hoosen E., Ehlers MM.

Genetic relatedness of Staphylococcus aureus isolates obtained from cystic fibrosis patients at a tertiary academic hospital in Pretoria, South Africa Scientific Reports 2018; 8: ArtNo: 12222

Malhotra S., Limoli DH., English AE., Parsek MR., Wozniak DJ.

Mixed Communities of Mucoid and Nonmucoid Pseudomonas aeruginosa Exhibit Enhanced Resistance to Host Antimicrobials mBio 2018; 9: 2:e00275-18

Mariappan V., Thavagnanam S., Vellasaamy KM., Teh CJS., Atiya N., Ponnampalavanar S., Vadivelu J.

Relapse of chronic melioidosis in a paediatric cystic fibrosis patient: first case report from Malaysia BMC Infectious Diseases 2018; 18: ArtNo: 455

Mastropasqua MC., Lamont I., Martin LW., Reid DW., D'Orazio A.

Pseudomonas aeruginosa to express virulence traits and colonize the human lung Molecular Microbiology 2018; 109: 1069


Molecular epidemiology of Pseudomonas aeruginosa in an unsegregated bronchiectasis cohort sharing hospital facilities with a cystic fibrosis cohort Thorax 2018; 73: 677 - 679


Anaerobic bacteria cultured from cystic fibrosis airways correlate to milder disease: a multisite study European Respiratory Journal 2018; 52: 1:1800242

Muthu V., Sehgal IS., Dhooria S., Aggarwal AN., Agarwal R.

Utility of recombinant Aspergillus fumigatus antigens in the diagnosis of allergic bronchopulmonary aspergillosis: A systematic review and diagnostic test accuracy meta-analysis Clinical and Experimental Allergy 2018; 48: 1107 - 1136

Muthu V., Sehgal IS., Dhooria S., Bal A., Agarwal R.

Allergic bronchopulmonary aspergillosis presenting as nephrotic syndrome due to secondary amyloidosis: Case report and systematic review of the literature Lung India 2018; 35: 332 - 335


Exophiala dermatitidis isolates from various sources: using alternative invertebrate host organisms (Caenorhabditis elegans and Galleria mellonella) to determine virulence Scientific Reports 2018; 8: ArtNo: 12747

Phan J., Gallagher T., Oliver A., England WE., Whiteson K.

Fermentation products in the cystic fibrosis airways induce aggregation and dormancy-associated expression profiles in a CF clinical isolate of Pseudomonas aeruginosa FEMS Microbiology Letters 2018; 365: 10:6082

Pinckova T., Paquin-Prouls D., Moll M., Flodstrom-Tullberg M., Hjelte L., Sandberg JK.

Severely Impaired Control of Bacterial Infections in a Patient With Cystic Fibrosis Defective in Mucosal-Associated Invariant T Cells Chest 2018; 153: E93 - E96

Pompilio A., Geminiani C., Bosco D., Rana R., Aceto A., Bucciarrelli T., Scotti L., Di Bonaventura G.

Electrochemically Synthesized Silver Nanoparticles Are Active Against Planktonic and Biofilm Cells of Pseudomonas aeruginosa and Other Cystic Fibrosis-Associated Bacterial Pathogens Frontiers in Microbiology 2018; 9: ArtNo: 1349

Poore TS., Virella-Lowell L., Guimbellot JS.

Potential pathogenicity of Inquilinus limosus in a pediatric patient with cystic fibrosis Pediatric Pulmonology 2018; 53: E21 - E23

Pradepsan S., Wark P.

Pseudomonas pharyngitis in a cystic fibrosis patient Respiriology Case Reports 2018; 6: 5:USNP e00325

Price EP., Viberg LT., Kidd TJ., Bell SC., Currie BJ., Sarovich EMS.

A review of the literature on the approach to the diagnosis of allergic bronchopulmonary aspergillosis: A systematic review and meta-analysis of allergic bronchopulmonary aspergillosis presenting as nephrotic syndrome due to secondary amyloidosis: Case report and systematic review of the literature Clinical and Experimental Allergy 2018; 48: 1107 - 1136

Price EP., Viberg LT., Kidd TJ., Bell SC., Currie BJ., Sarovich EMS.

A review of the literature on the approach to the diagnosis of allergic bronchopulmonary aspergillosis: A systematic review and meta-analysis of allergic bronchopulmonary aspergillosis presenting as nephrotic syndrome due to secondary amyloidosis: Case report and systematic review of the literature Clinical and Experimental Allergy 2018; 48: 1107 - 1136

Recio R., Borkotoky S., Murali A., Suchiang K., Mohanty SK., Busi S.

Attestation of quorum sensing controlled virulence factors and biofilm formation in Pseudomonas aeruginosa by pentacyclic triterpenes, betulin and betulinic acid Microbial Pathogenesis 2018; 118: 48 - 60

Reece E., Doyle S., Greally P., Renwick J., McClean S.

Aspergillus fumigatus Inhibits Pseudomonas aeruginosa in Co-culture: Implications of a Mutually Antagonistic Relationship on Virulence and Inflammation in the CF Airway Frontiers in Microbiology 2018; 9: ArtNo: 1205

Non-Tuberculous Mycobacteria multispecies biofilms in cystic fibrosis: development of an in vitro Mycobacterium abscessus and Pseudomonas aeruginosa dual species biofilm model

*International Journal of Medical Microbiology* 2018; 308: 413 - 423

Rodriguez-Sevilla G., Rigouts C., Vandeplassche E., Ostyn L., Mahillo-Fernandez I., Esteban J., Peremarch CFP., Coeney T., Crabbe A.

Influence of three-dimensional lung epithelial cells and interspecies interactions on antibiotic efficacy against Mycobacterium abscessus and Pseudomonas aeruginosa

*Pathogens and Disease* 2018; 76: 4; jfy034

Ronchetti K., Tame JD., Paisey C., Thia LP., Doull I., Howe R., Mahenthiralingam E., Forton JT.

The CF-Sputum Induction Trial (CF-SpIT) to assess lower airway bacterial sampling in young children with cystic fibrosis: a prospective internally controlled interventional trial

*Lancet Respiratory Medicine* 2018; 6: 461 - 471

Rossi E., Falcon M., Molin S., Johansen HK.

High-resolution in situ transcriptions of Pseudomonas aeruginosa unveils genotype independent patho-phenotypes in cystic fibrosis lungs

*Nature Communications* 2018; 9: ArtNo: 3459

Saeed A., Bosch A., Bettiol M., Gonzalez DLN., Erben MF., Lamberti Y.

Novel Guanine Compound against Multidrug-Resistant Cystic Fibrosis-Associated Bacterial Species

*Molecules* 2018; 23: S1158

Salloum T., Nassour E., Araj GF., Abhoud E., Tokajian S.

Insights into the genome diversity and virulence of two clinical isolates of Burkholderia cepacia cenocepacia

*Journal of Medical Microbiology* 2018; 67: 1157 - 1167


CF Lung Disease - a German S3 Guideline: Module 2: Diagnostics and Treatment in Chronic Infection with Pseudomonas aeruginosa

*Pneumonology* 2018; 72: 347 - 392


Prevalence and characterization of azole-resistant Aspergillus fumigatus in patients with cystic fibrosis: a prospective multicentre study in Germany

*Journal of Antimicrobial Chemotherapy* 2018; 73: 2047 - 2053

Sfeir MM.

Burkholderia cepacia complex infections: More complex than the bacterium name suggest

*Journal of Infection* 2018; 77: 166 - 170


RpoN-Dependent Direct Regulation of Quorum Sensing and the Type VI Secretion System in Pseudomonas aeruginosa PA01

*Journal of Bacteriology* 2018; 200: 16:e00205-18

Silva IN., Pessoa FD., Ramires MJ., Santos MR., Becker JD., Cooper VS., Moreira LM.

The OmpR Regulator of Burkholderia multivorans Controls Mucoid- to-Nonmucoid Transition and Other Cell Envelope Properties Associated with Persistence in the Cystic Fibrosis Lung

*Journal of Bacteriology* 2018; 200: 17-e00216-18


Molecular typing of Burkholderia cepacia complex isolated from patients attending an Italian Cystic Fibrosis Centre

*New Microbiologia* 2018; 41: 141 - 144

Tippett E., Ellis S., Wilson J., Kotsimbos T., Spelman D.

Mycobacterium abscessus Complex: Natural History and Treatment Outcomes at a Tertiary Adult Cystic Fibrosis Center


Tisot A., Thomas MF., Corris PA., Brodlie M.

Non-Tuberculous Mycobacteria infection and lung transplantation in cystic fibrosis: a worldwide survey of clinical practice

*BMC Pulmonary Medicine* 2018; 18: ArtNo: 86


Impact of T2R38 Receptor Polymorphisms on Pseudomonas aeruginosa Infection in Cystic Fibrosis

*American Journal of Respiratory and Critical Care Medicine* 2018; 197: 1635 - 1658

Van Dalem A., Herpel M., Echahidi F., Peeters C., Wybo I., De Wachter E., Vandamme P., Pierard D.

In Vitro Susceptibility of Burkholderia cepacia Complex Isolated from Cystic Fibrosis Patients to Ceftriaxime-Avibactam and Ceflofoxane-Tazobactam

*Antimicrobial Agents and Chemotherapy* 2018; 62: 9:e00590-18


Survival after lung transplantation of cystic fibrosis patients infected with Burkholderia dolosa (genomovar VI)

*Clinical Transplantation* 2018; 32: 5:e1326

Whee BA., Tai AS., Sherrard LJ., Ben Zakour NL., Hanks KR., Kidd TJ., Ramsay KA., Lamont I., Whiteley DM., Bell SC., Beaton SA.

Whole genome sequencing reveals the emergence of a Pseudomonas aeruginosa shared strain sub-lineage among patients treated within a single cystic fibrosis centre

*BMC Genomics* 2018; 19: ArtNo: 644

Williams D., Fothergill JL., Evans B., Caples J., Haldenby S., Walshaw MJ., Brochhurst MA., Winstanley C., Paterson S.

Transmission and lineage displacement drive rapid population genomic flux in cystic fibrosis airway infections of a Pseudomonas aeruginosa epidemic strain

*Microbial Genomics* 2018; 4: 3:00167

Zuttion F., Ligeour C., Vidal O., Walte M., Morvan F., Vidal S., Vassee JJ., Chevolot Y., Phaner-Goutorbe M., Schillers H.

The anti-adhesive effect of glycoclusters on Pseudomonas aeruginosa bacteria adhesion to epithelial cells studied by AFM single cell force spectroscopy

*Nanoscale* 2018; 10: 12771 - 12778

**Nutrition**


Nutritional Considerations in Pediatric Pancreatitis: A Position Paper from the NASPHAN Pancreas Committee and ESPHAN Cystic Fibrosis/Pancreas Working Group

*Journal of Pediatric Gastroenterology and Nutrition* 2018; 67: 131 - 143

de Assis MJC., Cartaxo CGB., Costa MJD., Queiroz DJM., Arruda MC., Goncalves MDR.

Association between hematological profile and serum 25-hydroxyvitamin D levels and Foxi polymorphism in individuals with cystic fibrosis

*Revista de Nutricao* 2018; 39: 2101 - 2105


Review on Vitamin K Deficiency and its Association with Hemostasis in Individuals with Cystic Fibrosis

*Clinical Transplantation* 2017; 31: 2101 - 2105

**de Assis MJC., Cartaxo CGB., Costa MJD., Queiroz DJM., Arruda MC., Goncalves MDR.**

Association between hematological profile and serum 25-hydroxyvitamin D levels and Foxi polymorphism in individuals with cystic fibrosis

*Revista de Nutricao* 2018; 39: 2101 - 2105


Review on Vitamin K Deficiency and its Association with Hemostasis in Individuals with Cystic Fibrosis

*Clinical Transplantation* 2017; 31: 2101 - 2105
Respiratory Care Technique in Subjects With Chronic Obstructive Airway Diseases

JMIR Mhealth and Uhealth 2018; 6: e113

Jessup M., Li A., Fulbrook P., Bell SC.
The experience of men and women with cystic fibrosis who have become a parent: A qualitative study
Journal of Clinical Nursing 2018; 27: 1702 - 1712

Keyte R., Egan H., Jackson C., Nash E., Regan A., Mantziou M.
A weekend/weekday comparison of adherence to daily treatment regimens in adults with cystic fibrosis
Health Psychology Report 2018; 6: 146 - 157

Knudsen KB., Boisen KA., Katzenstein TL., Mortensen LH., Pressler T., Skov M., Jarden M.
Living with cystic fibrosis - a qualitative study of a life coaching intervention
Patient Preference and Adherence 2018; 12: 585 - 594

Li SS., Hayes D., Tobias JD., Morgan WJ., Tumin D.
Health insurance and use of recommended routine care in adults with cystic fibrosis

Lonabaugh KP., O'Neal KS., McIntosh H., Condren M.
Cystic fibrosis-related education: Are we meeting patient and caregiver expectations?
Patient Education and Counseling 2018; 101: 1865 - 1870

Orenstein DM., Ahoud RN.
Cost(s) of caring for patients with cystic fibrosis
Current Opinion in Pediatrics 2018; 30: 393 - 398

Quigley SJ., Linnane B., Connellan S., Ward A., Ryan P.
Psychosocial Distress and Knowledge Deficiencies in Parents of Children in Ireland Who Caruy an Altered Cystic Fibrosis Gene
Journal of Genetic Counseling 2018; 27: 589 - 596

Wood J., Jenkins S., Putrino D., Mulrennan S., Morey S., Cecins N., Hill K.
High usability of a smartphone application for reporting symptoms in adults with cystic fibrosis
Journal of Telemedicine and Telecare 2018; 24: 398 - 402

Zubrzycka R.
Coping with stress by mothers of children and adolescents with cystic fibrosis
Advances in Respiratory Medicine 2018; 86: 86 - 91

Pulmonology

Aanaes K., Alanin MC., Nielsen KG., Jorgensen MM., van Buchwald C., Holby J., Johansen HK., Johannesen HH., Mortensen J.
The accessibility of topical treatment in the paranasal sinuses on operated cystic fibrosis patients assessed by scintigraphy
Rhinology 2018; 56: 268 - 273

Combret Y., Medrinal C., Prieur G., Quesada AR., Le Roux P., Reycher G.
Effect of backpack carrying on forced vital capacity in cystic fibrosis: A randomized crossover-controlled trial

De Rose V., Mollory K., Gohy S., Pilette C., Greene CM.
Airway Epithelium Dysfunction in Cystic Fibrosis and COPD
Mediators of Inflammation 2018; 1: 1309746

Intrapulmonary percussive ventilation improves lung function in cystic fibrosis patients chronically colonized with Pseudomonas aeruginosa: a pilot cross-over study
European Journal of Clinical Microbiology & Infectious Diseases 2018; 37: 1143 - 1151

El Basha N.
Impact of underlying cause of bronchiectasis on clinical outcome: A comparative study on CF and Non-CF bronchiectasis in Egyptian children
Egyptian Pediatric Association Gazette 2018; 66: 49 - 53

Gais T., Bregy L., Stehler N., Gaugt MM., Bruderer T., Garcia-Gomez D., Moeller A., Singer F., Schwarz EL., Benden C., Sinues PML., Zenobi R., Kohler M.
Real-time exhaled breath analysis in patients with cystic fibrosis and controls

Hilton N., Solis-Moya A.
Respiratory muscle training for cystic fibrosis (Review)
Cochrane Database of Systematic Reviews 2018: :5:CD006112

Insa K., Elisabeth K., Sophie Y., Nicolas R., Sylvia N., Kathryn R., Carmen C., Philipp L.
The Swiss Cystic Fibrosis Infant Lung Development (SCILD) cohort
Swiss Medical Weekly 2018; 148: ArtNo: w14618

Konig P., Ner Z., Acton JD., Ge B., Hewett J.
Is an FEV1 of 80% predicted a normal spirometry in cystic fibrosis children and adults?
Clinical Respiratory Journal 2018; 12: 2397 - 2403

Koucky V., Skalicka V., Pohnuek P.
Nitrogen multiple breath washout test for infants with cystic fibrosis
European Respiratory Journal 2018; 52: 2:1800815

The expression of Mirc1/Mir17-92 cluster in sputum samples correlates with pulmonary exacerbations in cystic fibrosis patients
Journal of Cystic Fibrosis 2018; 17: 454 - 461

Ma JT., Tang C., Kang L., Voynov JA., Rubin BK.
Cystic Fibrosis Sputum Rheology Correlates With Both Acute and Longitudinal Changes in Lung Function
Chest 2018; 154: 370 - 377

Pooling of bronchoalveolar lavage in children with cystic fibrosis does not adversely affect the microbiological yield or sensitivity in detecting pulmonary inflammation
Journal of Cystic Fibrosis 2018; 17: 391 - 399

Monroe EJ., Pierce DB., Ingraham CR., Johnson GE., Shivaram GM., Valji K.
An Interventionalist's Guide to Hemoptysis in Cystic Fibrosis
Radiographics 2018; 38: 624 - 641

Pallin M., Keating D., Kaye DM., Kotsumbos T., Wilson JW.
Subclinical Left Ventricular Dysfunction is Influenced by Genotype Severity in Patients with Cystic Fibrosis
Clinical Medicine Insights-circulatory Respiratory and Pulmonary Medicine 2018; 12: ArtNo: UNSP 117

Pezoa A., Jarquera P., Madrid R., Maturana P., Viviani P., Causade S.
Spirometric characterization of cystic fibrosis patients
Revista Chilena de Pediatría-chile 2018; 89: 312 - 338

Radtkle T., Boni L., Bohnacker P., Fischer P., Bendenc D., Dressel H.
The many ways sputum flows - Dealing with high within-subject variability in cystic fibrosis sputum rheology
Respiratory Physiology & Neurobiology 2018; 254: 36 - 39

Ring AM., Buchvald FF., Holgersen MG., Green K., Nielsen KG.
Fitness and lung function in children with primary ciliary dyskinesia and cystic fibrosis
Respiratory Medicine 2018; 139: 79 - 85
McCarthy C., Brewington JJ., Harkness B., Clancy JP., Trapnell BC.
Personalised CFTR pharmacotherapeutic response testing and therapy of cystic fibrosis
European Respiratory Journal 2018; 51: 6:1702457

McCrey BE., Harper HN., McPhail GL.
Use and Incidence of Adverse Effects of Proton Pump Inhibitors in Patients with Cystic Fibrosis
Pharmacotherapy 2018; 38: 725 - 729

Moore PJ., Taran R.
The epithelial sodium channel (ENaC) as a therapeutic target for cystic fibrosis lung disease
Expert Opinion on Therapeutic Targets 2018; 22: 687 - 701

Nemchenko AS., Panfilova AL., Podgaina MV., Zaytzeva YL., Balynska MV.
Pharmacoeconomic Assessment of the Application of Domslave Alfa in the Treatment of Lung Generation in Patients with Cystic Fibrosis
Asian Journal of Pharmaceutics 2018; 12:

Ratjen F., Klingel M., Black P., Powers MR., Grasemann H., Solomon M., Sagel SD., Donaldson SH., Rowe SM., Rosenfeld M.
Changes in Lung Clearance Index in Preschool-aged Patients with Cystic Fibrosis Treated with Ivcnafor (GOAL): A Clinical Trial
American Journal of Respiratory and Critical Care Medicine 2018; 198: 526 - 528

Robinson E., MacDonald KD., Slaughter K., McKinney M., Patel S., Sun C., Sahay G.
Lipid Nanoparticle-Delivered Chemically Modified mRNA Restores Chloride Secretion in Cystic Fibrosis
Molecular Therapy 2018; 26: 2034 - 2046

Rosenfeld M., Wainwright CE., Higgins M., Wang LT., Mckee C., Campbell D., Tian S., Schneider J., Cunningham S., Davies JC.
Ivcnafor treatment of cystic fibrosis in children aged 12 to < 24 months and with a CFTR gating mutation (ARIVAL): a phase 3 single-arm study
Lancet Respiratory Medicine 2018; 6: 545 - 553

Rubin BK.
Unmet needs in cystic fibrosis
Expert Opinion on Biological Therapy 2018; 18:

Segall M.
DEADLOCK OVER CYSTIC FIBROSIS DRUG Allocating healthcare resources-seriously ill people should have priority
British Medical Journal 2018; 361: ArtNo: k1817

Shah GB., De Keyzer L., Russell JA., Halderman A.
Treatment of chronic rhinosinusitis with dornase alfa in patients with cystic fibrosis: a systematic review
International Forum of Allergy & Rhinology 2018; 8: 729 - 736

Strug LJ., Stephenson AL., Panjwani N., Harris A.
Recent advances in developing therapeutics for cystic fibrosis
Human Molecular Genetics 2018; 27: R173 - R186

Toseo A., Villella VR., Castaldo A., Kroemer G., Maiuri L., Raia V.
Repurposing therapies for the personalised treatment of cystic fibrosis
Expert Opinion on Orphan Drugs 2018; 6: 361 - 373

Van Biervliet S., Hauser B., Verhulst S., Stepman H., Delanghe J., Warzee JP., Pot B., Vandeweile T., Wilschanski M.
Probiotics in cystic fibrosis patients: A double blind crossover placebo controlled study Pilot study from the ESPGHAN Working Group on Pancreas/CF
Clinical Nutrition Espen 2018; 27: 59 - 65

Vasudevan N., Sharma MK., Reddy DS., Kulkarni AA.
A multi-step continuous flow synthesis of the cystic fibrosis medicine ivacaftor
Reaction Chemistry & Engineering 2018; 3: 520 - 526

Transplantation
Borchi B., Ocampo MB., Cimino G., Pizzaniglia G., Bresl S., Braggion C.
Mortality rate of patients with cystic fibrosis on the waiting list and within one year after lung transplantation: a survey of Italian CF centers
Italian Journal of Pediatrics 2018; 44: ArtNo: 72

Ho C., Hayes D., Khosravi M., Spleagard ML., Tumin D., Lloyd EA.
Sedation with Prophol for Bronchoscopy in Cystic Fibrosis Lung Transplant Recipients
Lung 2018; 196: 435 - 439

Jardel S., Reynaud Q., Durieu I.
Long-term extrapulmonary comorbidities after lung transplantation in cystic fibrosis: Update of specificities
Clinical Transplantation 2018; 32: 6:e13269

Jauregui A., Deu M., Romero L., Roman A., Moreno A., Armengol M., Sole J.
Lung Transplantation in Cystic Fibrosis and the Impact of Extracorporeal Circulation
Archivos de Bronconeumologia 2018; 54: 313 - 319

Kounis L., Levy P., Rebours V.
Ivcnafor CFTR Potentiator Therapy is Efficient for Pancreatic Manifestations in Cystic Fibrosis
American Journal of Gastroenterology 2018; 113: 1058 - 1059

Orfanos S., Gomez C., Baron S., Akkisetty R., Dufeu N., Coltey B., Thomas PA., Rolain JM., Reynaud-Gautbert M.
Impact of gram negative bacteria airway recolonization on the occurrence of chronic lung allograft dysfunction after lung transplantation in a population of cystic fibrosis patients
BMC Microbiology 2018; 18: ArtNo: 88

Saldanha LJ., Akinneyde O., Robinson KA.
Immunosuppressive drug therapy for preventing rejection following lung transplantation in cystic fibrosis
Cochrane Database of Systematic Reviews 2018; : 6:CD009421

Staufe K., Halibasic E., Hillebrand P., Harm S., Schwarz S., Jaksch P., Kivaranovic D., Klepetko W., Trauner M., Kazemi-Shirazi L.
Impact of nutritional status on pulmonary function after lung transplantation for cystic fibrosis
United European Gastroenterology Journal 2018; 6: 1049 - 1055

Urology
Nowakowski ACH.
Cystic Fibrosis Kidney Disease: 10 Tips for Clinicians
Frontiers in Medicine 2018; 5; ArtNo: 242