

## 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](http://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

41<sup>st</sup> European Cystic Fibrosis Conference, Belgrade, Serbia, 8<sup>th</sup> 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
- 16<sup>th</sup> ECFS Basic Science Conference, Dubrovnik 27-30 March 2019
- Young Investigators Meeting, Paris 27 Feb./01 March 2019
- 42<sup>nd</sup> 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 preminent 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](mailto: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

-Best practice: ECFS Book proposal on CF pharmacopoeia

-Guideline for guidelines

-Cooperation with ERN-LUNG

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

-Increase coverage to reach 80% or higher in countries participating to the ECFSPR.

-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 - 3<sup>rd</sup> 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 27<sup>th</sup>.

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



### Denver, CO, US

*17 October 2018:*

ECFS Board Meeting

*18-20 October 2018:*

33rd North American Cystic Fibrosis Conference

### Brussels, Belgium

*23 January 2019:* ECFS Winter Board Meeting

*24 January 2019:* ECFS Standards of Care Meeting

*24-25 January 2019:*

ECFS CTN Steering Group Meeting

*25 January 2019:*

ECFS Patient Registry Steering Group Meeting

### Tunis, Tunisia

*14-16 February 2019*

ECFS Diagnostic Network Working Group Meeting

### Paris, France

*27-February - 01 March 2019*

13<sup>th</sup> European Young Investigators Meeting

Abstract submission from 01 October to 04 November 2018

### Liverpool, United Kingdom

*04 June 2019:*

ECFS Board Meeting

*05-08 June 2019:*

42<sup>nd</sup> European CF Conference

### Dubrovnik, Croatia

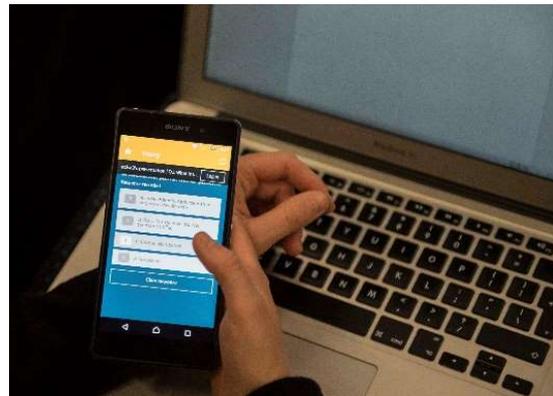
*27-30 March 2019*

ECFS 16<sup>th</sup> 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.

## CF Reference List

(compiled between May & September 2018)

### Adults & Adolescents

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Descriptions of the Pain Experience in Adults and Adolescents with Cystic Fibrosis

*Pain Management Nursing* 2018; 19: 340 - 347

**Bernstein RM., Riekert KA., Quittner AL.**

Measuring Knowledge of Disease Management in Adolescents with Cystic Fibrosis: Initial Psychometric Evaluation

*Pediatric Allergy Immunology and Pulmonology* 2018; 9: ArtNo: 306

**Chatterjee K., Goyal A., Reddy D., Koppurapu V., Innabi A., Alzghoul B., Jagana R.**

Prevalence and predictors of readmissions among adults with cystic fibrosis in the United States

*Advances in Respiratory Medicine* 2018; 86: 75 - 77

**Durieu I., Reynaud Q., Lega JC.**

Transition: From pediatric to adult care center

*Revue de Medecine Interne* 2018; 39: A45 - A47

**Kazmerski TM., Hill K., Prushinskaya O., Nelson E., Greenberg J., Pitts SAB., Borrero S., Miller E., Sawicki GS.**

Perspectives of adolescent girls with cystic fibrosis and parents on disease-specific sexual and reproductive health education

*Pediatric Pulmonology* 2018; 53: 1027 - 1034

**Nap-van der Vlist MM., Burghard M., Hulzebos HJ., Doeleman WR., Heijerman HGM., van der Ent CK., Nijhof SL.**

Prevalence of severe fatigue among adults with cystic fibrosis: A single center study

*Journal of Cystic Fibrosis* 2018; 17: 368 - 374

**Rousset-Jablonski C., Reynaud Q., Nove-Josserand R., Durupt S., Durieu I.**

Gynecological management and follow-up in women with cystic fibrosis

*Revue des Maladies Respiratoires* 2018; 35: 592 - 603

**Saez-Flores E., Tonarely NA., Barker DH., Quittner AL.**

Examining the Stability of the Hospital Anxiety and Depression Scale Factor Structure in Adolescents and Young Adults With Cystic Fibrosis: A Confirmatory Factor Analysis

*Journal of Pediatric Psychology* 2018; 43: 625 - 635

**Seller EP., Juan PM.**

Analysis of social and work reality of adult people with Cystic Fibrosis disease: The situation of the Murcia Region (Spain)

*Interaccion Y Perspectiva* 2018; 8: 47 - 66

**Stevens D., Neyedli HF.**

Clinical Model of Exercise-Related Dyspnea in Adult Patients With Cystic Fibrosis

*Journal of Cardiopulmonary Rehabilitation and Prevention* 2018; 38: 187 - 192

### Animal Model

**Hahn A., Salomon JJ., Leitz D., Feigenbutz D., Korsch L., Lisewski I., Schrimpf K., Millar-Buchner P., Mall MA., Frings S., Mohrlen F.**

Expression and function of Anoctamin 1/TMEM16A calcium-activated chloride channels in airways of in vivo mouse models for cystic fibrosis research

*Pflugers Archiv-European Journal of Physiology* 2018; 470: 1335 - 1348

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A G542X cystic fibrosis mouse model for examining nonsense mutation directed therapies

*PLoS One* 2018; 13: e0199573

**Meyerholz DK., Stoltz DA., Gansemer ND., Ernst SE., Cook DP., Strub MD., LeClair EN., Barker CK., Adam RJ., Leidinger MR., Gibson-Corley KN., Karp PH., Welsh MJ., McCray PB.**

Lack of cystic fibrosis transmembrane conductance regulator disrupts fetal airway development in pigs

*Laboratory Investigation* 2018; 98: 825 - 838

**Philp AR., Riquelme TT., Millar-Buchner P., Gonzalez R., Sepulveda FV., Cid LP., Flores CA.**

Kcnn4 is a modifier gene of intestinal cystic fibrosis preventing lethality in the Cftr-F508del mouse

*Scientific Reports* 2018; 8: ArtNo: 9320

**Portal C., Gouyer V., Leonard R., Husson MO., Gottrand F., Desseyn JL.**

Long-term dietary (n-3) polyunsaturated fatty acids show benefits to the lungs of CFTR F508del mice

*PLoS One* 2018; 13: e0197808

**Rosen BH., Chanson M., Gawenis LR., Liu JH., Sofoluwe A., Zoso A., Engelhardt JF.**

Animal and model systems for studying cystic fibrosis

*Journal of Cystic Fibrosis* 2018; 17:

**Rosen BH., Evans TIA., Moll SR., Gray JS., Liang B., Sun XS., Zhang YL., Jensen-Cody CW., Swatek AM., Zhou WH., He N., Roti PG., Tyler SR., Keiser NW., Anderson PJ., Brooks L., Li YL., Pope RM., Rajput M., Hoffman EA., Wang K.**

Infection Is Not Required for Mucoinflammatory Lung Disease in CFTR-Knockout Ferrets

*American Journal of Respiratory and Critical Care Medicine* 2018; 197: 1308 - 1318

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Inhibition of *Pseudomonas aeruginosa* secreted virulence factors reduces lung inflammation in CF mice

*Virulence* 2018; 9: 1008 - 1018

### Antimicrobials

**Ahmed MI., Mukherjee S.**

Treatment for chronic methicillin-sensitive *Staphylococcus aureus* pulmonary infection in people with cystic fibrosis (Review)

*Cochrane Database of Systematic Reviews* 2018; : 7:CD011581

**Al-Nemrawi NK., Alshraideh NAH., Zayed AL., Altaani BM.**

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