

ECFS NEWSLETTER - Issue 56 - January 2018

01. Letter from the President
02. Upcoming Events
03. Deadlines
04. ECFS Award - Call for nominations
05. Gerd Döring Award - Call for nominations
06. ECFS Board Elections - Call for nominations
07. ECFS Meetings - CTN, Patient Registry, Standards of Care
08. HIT-CF project launched
09. ECFS Strategic Plan for new treatments
10. In memoriam Lutz Goldbeck
11. 41st European CF Conference - Belgrade
12. Belgrade Conference reduced registration fee
13. Nutrition Masterclass
14. Physiotherapy Short Course - Airway Clearance Techniques
15. An introductory course in Cognitive Behavioral Therapy and Interpersonal Therapy
16. CF course - the treatment of adults with CF - an in-depth course
17. Current References in CF

European Cystic Fibrosis Society
 Kastanieparken 7
 7470 Karup, Denmark
 Tel: +45 86 676260
 Fax: +45 86 676290
info@ecfs.eu
www.ecfs.eu

01. Letter from the President

Dear Friends,

I hope you had a good start of the year. I wish you and your family all the best for 2018!

2017 was another busy year for the European Cystic Fibrosis Society, with not only the Basic Science Conference in Portugal and the Annual conference in Seville, but a number of special projects and Working Group meetings were held and some selected updates are given below.



There will be ECFS Board elections in 2018. Changes in the ECFS Board are already happening. Isabelle Fajac will take on the role of President and will officially start her 3-year mandate at the ECFS conference this June. Also, Harm Tiddens and Daniel Peckham will end their terms in June.

I would like to encourage you all to consider your own nomination or to nominate an ECFS member you believe would benefit the ECFS community by being actively engaged in the Board. Please consider how important these elections are for the future of the Society. Further information about the elections is included later in the Newsletter.

Planning for the 2018 Conference is well underway with all necessary information available on the website. I would like to thank all those who have submitted abstracts and all the speakers who have accepted to contribute to the conference. This year, we introduced a special discounted rate for attendees from lower income countries in an effort to encourage participation of healthcare professionals from these countries. We have excellent programmes and courses planned and we look forward to your participation.

We have a lot to do over the coming months and I hope you will join us for some of these activities.

Many thanks, as always, to Dr. Henry Ryley for compiling the current references in cystic Fibrosis contained in this Newsletter

Please contact us if you have news items you would like to have included in future Newsletters or published on our website.

Yours sincerely,

Kris De Boeck, ECFS President

02. Upcoming Events

- ECFS Diagnostic Network Working Group Meeting - 08-10 February 2018, St-Gallen, Switzerland
- 12th European Young Investigators Meeting - 21-23-February 2018, Paris, France
- 15th ECFS Basic Science Conference - 21-24 March 2018, Loutraki, Greece
- ECFS Board Elections
- ECFS Board Meeting - 05 June 2018, Belgrade, Serbia
- 41st European CF Conference - 06-09 June 2018, Belgrade, Serbia



03. Deadlines

- | | |
|----------------------------------|------------------|
| - Nomination ECFS Award | 22 February 2018 |
| - Nomination Gerd Döring Award | 22 February 2018 |
| - Nomination ECFS Board Election | 15 March 2018 |

04. ECFS Award - Call for nominations

The ECFS Award is given annually to honour a person who has made an outstanding contribution to our basic understanding of cystic fibrosis or to the treatment or care of patients with cystic fibrosis. The winner of the award will be invited to present a lecture at the Opening Plenary on 6 June 2018 of the annual conference in Belgrade.

You are cordially invited to nominate a candidate for this award. The deadline for proposals is **22 February 2018**. Please mail your proposal, accompanied by a detailed motivation and curriculum vitae of the candidate to the ECFS Executive Director Christine Dubois (christine.dubois@ecfs.eu).

05. Gerd Döring Award - Call for nominations

The Gerd Döring Award is a recent initiative of the European Cystic Fibrosis Society and is given annually to honour an exceptional early career young European scientist. The Award, a monetary donation of 5,000 euro to support research, will be presented at the Opening Plenary of the annual ECFS conference in Belgrade.

The award will be judged primarily on a paper published in the previous 3 calendar years (2015-2017) which has made a significant impact on the understanding or treatment of Cystic Fibrosis. Personal motivation and CV will make a 20% contribution to the scoring.

The award is open to PhD students and post-doctoral researchers with up to a maximum of four years' academic research experience following the completion of their PhD, or be of equivalent professional standing at the date of publication of the paper.

We encourage mentors, supervisors and co-workers of today's most exceptional early career European scientists to send in nominations for this competition. Self-nomination is also encouraged. The deadline for proposals is **February 22, 2018**. Please mail your proposal, accompanied by a detailed motivation, a PDF of the nominated paper and curriculum vitae of the candidate to the ECFS Executive Director Christine Dubois (christine.dubois@ecfs.eu)

06. ECFS Board Elections - Call for nominations



The ECFS cordially invites nominations for the following Board positions.

3 Board Positions: Harm Tiddens and Daniel Peckham will finish their terms on the Board in 2018.

Isabelle Fajac will become ECFS President in June and ends her mandate as Board member.

Job Descriptions and person specifics are available for Board members. [Board Member's Job Description](#).

Nominations should be sent to the ECFS Executive Director Christine Dubois (christine.dubois@ecfs.eu) by **15 March 2018** together with a motivational statement, confirmation that the candidate has agreed to the nomination, and a curriculum vitae. All nominated candidates must be current members of the ECFS. Information about the nominated candidates will be sent to the membership in April for an online vote prior to the Annual General Meeting in June where the results will be announced.

07. ECFS Meetings - CTN, Patient Registry, Standards of Care



The ECFS Clinical Trials Network, the ECFS Patient Registry Steering Committees met on 25-26 January in Brussels for their annual winter meetings. The Standards of Care Group also met in Brussels. This was an excellent opportunity to meet across ECFS projects, discuss common subjects and find new and even better opportunities for cooperation.

08. HIT-CF project launched!

In January 2018 the HIT-CF project kicked off.

This European project brings together researchers, doctors, pharmaceutical companies and patient representatives with the aim to develop 'personalized treatments' for Cystic Fibrosis (CF) patients throughout Europe and ultra-rare genetic profiles. The project is funded by the EU under the Horizon 2020 framework.

Check the video presentation [here](#).

Press release available [here](#)

09. ECFS Strategic Plan for new treatments

The unprecedented progress over the last few years in the development of new drugs targeting the cystic fibrosis basic defect whilst extremely encouraging, presents unique challenges. There are substantial disparities around the globe in patient access to both trials and licensed drugs and concerns over how future drugs will best be tested in the congested space of a small disease population. Patients with the rarest of mutations, which are often under-researched and poorly understood, may be particularly disadvantaged.

Within the ECFS, we have brought together a small group tasked with developing a ‘Strategic Plan’ to maximise progress in research as well as access to new, effective treatments.

Jane Davies leads the group, and a first workshop has been organised in Brussels in December 2017. The aim was to tackle the challenges from multiple angles and therefore the meeting brought together representatives from the ECFS, CTN clinical trials teams, international patient organisations, registries, trial design and health economics groups, and members of pharmaceutical and regulatory agencies.

The discussions were very fruitful and we will most probably organise a second meeting with the group at the conference in Belgrade

10. Obituary - Lutz Goldbeck, 25.11.1958 - 30.10.2017

In the fall of 2017 we unexpectedly lost Lutz Goldbeck, one of our lead psychologists in CF. Our heart goes out to the family that is confronted with such a sudden loss



Since 1985, Lutz Goldbeck has worked as a clinical psychologist and researcher across the disciplines of child psychiatry, psychotherapy and pediatrics. His research concentrated on psychological adaptation to chronic diseases, health-related quality of life, psychotraumatology, and the development and evaluation of psychosocial interventions including mental health.

He was the director of a state-certified training program for child and adolescent psychotherapists. He has participated in multiple interdisciplinary research collaborations funded e.g. by the EU, the Federal German Ministry of Health and the Federal Ministry of Education and Research (BMBF). From the beginning of his professional career he was dedicated to cystic fibrosis. Despite a wide diversity of other research topics he always had the topic of living with a chronic disease such as CF in mind. He represented the German community in international collaborations and helped to implement psychological care in German CF Clinics. He was one of the founding members of the Mental Health Working Group of the ECFS. His discreet and earnest temperament and style of argumentation was always a benefit for the whole group. Working with him has been a great pleasure for all of us. We will miss him sorely.

11. 41st European CF Conference - Belgrade, Serbia

Every year the Annual Conference travels through Europe landing to another destination. Organising the conference in one country also means to bring awareness to the local authorities about CF and all the challenges for patients and research.

We definitely think that choosing Belgrade to host our conference will make a significant impact on the whole region and will help the local CF community in their discussions with the Health Authorities and raise awareness about Cystic Fibrosis.

With your participation to the conference, you will contribute to make it a resounding success and we are sure you will also enjoy discovering sunny Belgrade.

The city lies on the confluence of two major European rivers, Sava and Danube, giving the city a very special charm.

Belgrade is flourishing with a lively artistic and cultural scene. Be ready to discover a fascinating, vibrant city, with some surprisingly attractive pedestrianised streets in the centre, and with a lot of charming places along the riverbanks.

Not registered yet? Please find all details [here](#)



12. Conference reduced registration rate

This year we introduced a reduced registration rate for participants working in countries classified as low-income economies, lower-middle-income economies and upper-middle-income economies by the World Bank. We hope this opportunity will encourage many more healthcare professionals to attend the conference. More information can be found [here](#).

13. Nutrition masterclass - practical management of nutrition issues in CF

Tuesday 5 June - Half day

This nutrition masterclass focuses on the practical aspects of assessing, monitoring and optimizing the nutritional status of Cystic fibrosis (CF) patients specific to the low- and middle-income economies healthcare setting. It is designed for all healthcare providers who have interests in the nutritional aspects of CF or who are new to CF. The aim will be to share effective nutritional management strategies where time, health care infrastructure and finance may be limited to optimize CF outcomes.

Topics will include: nutritional assessment (anthropometrics and dietary history), optimizing enzyme therapy, increasing calories/supplemental nutritional therapy, managing difficult feeding behaviours and effective treatment of early poor growth.

The course will be supported by experienced CF nutrition experts from across Europe and at the end of the course attendees will receive a certificate of attendance.

[Registration information](#)

14. Physiotherapy short course - Airway Clearance Techniques: basic approach

Tuesday 5 June & Wednesday 6 June - One and a half days

This course is intended for physiotherapists, physical or respiratory therapists who require basic training in the physiological basis, practical application of ACT therapies, including inhalation. This course will be a stimulating and interactive teaching approach to airway clearance techniques and how to go from research interpretation to daily working practice recommendations.

[Registration information](#)



15. An introductory course in Cognitive Behavioral Therapy and Interpersonal Therapy

Wednesday 06 June 2018 - All day

The Mental Health guidelines recommend offering Cognitive Behavioral Therapy (CBT) and/or Interpersonal Therapy (IPT) to patients and parent caregivers when screening indicates moderate to severe symptoms of anxiety and/or depression.

CBT is a widely-used, evidenced-based, psychosocial intervention for improving mental health. It focuses on the development of personal coping strategies, problem solving and changing unhelpful patterns in cognitions (e.g. thoughts, beliefs, and attitudes), behaviors, and emotional regulation. IPT is a short-term treatment that encourages patients to regain control of mood and functioning. It is based on a treatment alliance in which the therapist empathically engages the patient, helps the patient feel understood and structures success experiences.

This one-day course at the initiative of the ECFS Mental Health Working Group consists of a basic introduction in CBT and IPT. The course will highlight the principles of assessment and a variety of intervention techniques. CBT and IPT have been applied in medical settings for decades and the course will include examples of CBT and IPT approaches in CF.

The course will be given by Alexandra Klein Raphaeli. Alexandra received her doctorate in Clinical Psychology from Yeshiva University, New York City (USA). She also holds a masters degree in Education from Teachers College, Columbia University, New York City (USA). Currently, Dr. Raphaeli is an instructor and specialty CBT/IPT supervisor at Tel Aviv University's University Counseling Center, and maintains a private practice in Israel. She has worked as a therapist in hospital settings, counseling centers, and within research trials, and has served as a supervisor for psychologists, psychiatrists, and social workers.

[Registration information](#)

16. CF course - the treatment of adults with CF - an in-depth course

Wednesday 6 June - All day

Each year, we organise a CF Course prior to the Conference. This year, the course will focus on the treatment of adults with CF. In many countries, there are now more adults than children with CF. The course is specifically designed for pulmonologists and will invite them to deepen their knowledge on the treatment of adults with CF.

[Registration information](#)

17. Current references in CF

Please scroll to next page for full list

CF Reference List

Adults & Adolescents

Dowaikh H., Morfin-Sherpa F., Reix P.

Acute chest pain in an adolescent with cystic fibrosis in September: Would you have thought about this?
Pediatric Pulmonology 2017; 52: E70 - E72

Faricy LE., Church G.

Sepsis and acute respiratory distress syndrome requiring extracorporeal life support in an adolescent with mild cystic fibrosis

Respiratory Medicine Case Reports 2017; 22: 235 - 237

Goralski JL., Nasr SZ., Uluer A.

Overcoming barriers to a successful transition from pediatric to adult care
Pediatric Pulmonology 2017; 52:

Helms SW., Christon LM., Dellen EP., Prinstein MJ.

Patient and Provider Perspectives on Communication About Body Image with Adolescents and Young Adults With Cystic Fibrosis

Journal of Pediatric Psychology 2017; 42: 1040 - 1050

Kazmerski TM., Borrero S., Sawicki GS., Abebe KZ., Jones KA., Tuchman LK., Weiner DJ., Pilewski JM., Orenstein DM., Miller E.

Provider Attitudes and Practices toward Sexual and Reproductive Health Care for Young Women with Cystic Fibrosis
Journal of Pediatric and Adolescent Gynecology 2017; 30: 546 - 552

Kerr H., Price J., Nicholl H., O'Halloran P.

Transition from children's to adult services for young adults with life-limiting conditions: A realist review of the literature
International Journal of Nursing Studies 2017; 76: 1 - 27

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

Coach to cope: feasibility of a life coaching program for young adults with cystic fibrosis
Patient Preference and Adherence 2017; 11: 1613 - 1623

Oud L.

Critical illness among adults with cystic fibrosis in Texas, 2004-2013: Patterns of ICU utilization, characteristics, and outcomes
PLoS One 2017; 12: 10:e0186770

Prados C., Lerin M., Cabanillas JJ., Gomez-Carrera L., Alvarez-Sala R.

How are the ancient cystic fibrosis patients? Cystic fibrosis diagnosed over 60 years-old

Respiratory Medicine Case Reports 2017; 21: 49 - 51

Animal Model

Adam RJ., Abou Alaiwa MH., Bouzek DC., Cook DP., Gansemer ND., Taft PJ., Powers LS., Stroik MR., Hoegger MJ., McMenimen JD., Hoffman EA., Zabner J., Welsh MJ., Meyerholz DK., Stoltz DA.

Postnatal airway growth in cystic fibrosis piglets

Journal of Applied Physiology 2017; 123: 526 - 533

Birket SE., Davis JM., Fernandez CM., Tuggle KL., Oden AM., Chu KK., Tearney GJ., Fanucchi MV., Sorscher EJ., Rowe SM.

Development of an airway mucus defect in the cystic fibrosis rat
JCI Insight 2018; 3: 1:e97199

Li DF., Wang J., Sun DJ., Gong XF., Jiang H., Shu JZ., Wang ZY., Long Z., Chen YG., Zhang ZL., Yuan L., Guan RJ., Liang X., Li ZY., Yao HW., Zhong NS., Lu WJ.

Tanshinone IIA sulfonate protects against cigarette smoke-induced COPD and down-regulation of CFTR in mice

Scientific Reports 2018; 8: ArNo: 376

Luan XJ., Belev G., Tam JS., Jagadeeshan S., Hassan N., Gioino P., Grishchenko N., Huang YY., Carmalt JL., Duke T., Jones T., Monson B., Burmester M., Simovich T., Yilmaz O., Campanucci VA., Machen TE., Chapman LD., Ianowski JP.

Cystic fibrosis swine fail to secrete airway surface liquid in response to inhalation of pathogens
Nature Communications 2017; 8: ArNo: 786

Stalvey MS., Havasi V., Tuggle KL., Wang DZ., Birket S., Rowe SM., Sorscher EJ.

Reduced bone length, growth plate thickness, bone content, and IGF-I as a model for poor growth in the CFTR-deficient rat
PLoS One 2017; 12: 11:e0188497

Tipirneni KE., Cho DY., Skinner DF., Zhang SY., Mackey C., Lim DJ., Woodworth BA.

Characterization of Primary Rat Nasal Epithelial Cultures in CFTR Knockout Rats as a Model for CF Sinus Disease
Laryngoscope 2017; 127: E384 - E391

Antimicrobials

AbdulWahab A., Zahraldin K., Ahmed MAS., Abu Jarir S., Muneer M., Mohamed SF., Hamid JM., Hassan AAI., Ibrahim EB.

The emergence of multidrug-resistant *Pseudomonas aeruginosa* in cystic fibrosis patients on inhaled antibiotics
Lung India 2017; 34: 527 - 531

Ahn Y., Kim JM., Lee YJ., LiPuma JJ., Hussong D., Marasa BS., Cerniglia CE.

Effects of Extended Storage of Chlorhexidine Gluconate and Benzalkonium Chloride Solutions on the Viability of *Burkholderia Cenocepacia*
Journal of Microbiology and Biotechnology 2017; 27: 2211 - 2220

Babin BM., Atangcho L., van Eldijk MB., Sweredoski MJ., Moradian A., Hess S., Tolker-Nielsen T., Newman DK., Tirrell DA.

Selective Proteomic Analysis of Antibiotic-Tolerant Cellular Subpopulations in *Pseudomonas Aeruginosa* Biofilms
mBio 2017; 8: 5:e01593-17

Banaschewski BJH., Baer B., Arsenault C., Jazey T., Veldhuizen EJA., Delport J., Gooyers T., Lewis JF., Haagsman HP., Veldhuizen RAW., Yamashita C.

The Antibacterial and Anti-inflammatory Activity of Chicken

Cathelicidin-2 combined with Exogenous Surfactant for the

Treatment of Cystic Fibrosis-Associated Pathogens

Scientific Reports 2017; 7: ArNo: 15545

Banerjee M., Moulick S., Bhattacharya KK., Parai D., Chattopadhyay S., Mukherjee SK.

Attenuation of *Pseudomonas aeruginosa* quorum sensing, virulence and biofilm formation by extracts of Andrographis paniculata

Microbial Pathogenesis 2017; 113: 85 - 93

Bensman TJ., Wang JS., Jayne J., Fukushima L., Rao AP., D'Argenio DZ., Beringer PM.

Pharmacokinetic-Pharmacodynamic Target Attainment Analyses to Determine Optimal Dosing of Ceftazidime-Avibactam for the Treatment of Acute Pulmonary Exacerbations in Patients with Cystic Fibrosis

Antimicrobial Agents and Chemotherapy 2017; 61: 10:e00988-17

Bos AC., Mouton JW., van Westreenen M., Andrinopoulou ER., Janssens HM., Tiddens HAWM.

Patient-specific modelling of regional tobramycin concentration levels in airways of patients with cystic fibrosis: can we dose once daily?

Journal of Antimicrobial Chemotherapy 2017; 72: 3435 - 3442

- Brockman SM., Bodas M., Silverberg D., Sharma A., Vij N.**
Dendrimer-based selective autophagy-induction rescues Delta F508-CFTR and inhibits *Pseudomonas aeruginosa* infection in cystic fibrosis
PLoS One 2017; 12: 9:pone0184793
- Da Costa L., Scheers E., Coluccia A., Rosetti A., Roche M., Neyts J., Terme T., Cirilli R., Mirabelli C., Silvestri R., Vanelle P.**
Heterocyclic pharmacocchemistry of new rhinovirus antiviral agents: A combined computational and experimental study
European Journal of Medicinal Chemistry 2017; 140: 528 - 541
- Dentice RL., Elkins MR., Dwyer GM., Bye PTP.**
The use of an alternate side lying positioning strategy during inhalation therapy does not prolong nebulisation time in adults with Cystic Fibrosis: a randomised crossover trial
BMC Pulmonary Medicine 2018; 18: ArNo: 3
- Diez-Aguilar M., Morosini MI., Koksal E., Oliver A., Ekkelenkamp M., Canton R.**
Use of Calgary and Microfluidic BioFlux Systems to Test the Activity of Fosfomycin and Tobramycin Alone and in Combination against Cystic Fibrosis *Pseudomonas Aeruginosa* Biofilms
Antimicrobial Agents and Chemotherapy 2018; 62: 1:e01650-17
- El-Halfawy OM., Naguib MM., Valvano MA.**
Novel antibiotic combinations proposed for treatment of Burkholderia cepacia complex infections
Antimicrobial Resistance and Infection Control 2017; 6: ArNo: 120
- Firsov AA., Alieva KN., Strukova EN., Golikova MV., Portnoy YA., Dovzhenko SA., Kobrin MB., Romanov AV., Edelstein MV., Zinner SH.**
Testing the mutant selection window hypothesis with *Staphylococcus aureus* exposed to linezolid in an in vitro dynamic model
Journal of Antimicrobial Chemotherapy 2017; 72: 3100 - 3107
- Forier K., Van Heck V., Carlier M., Van Braeckel E., Van Daele S., De Baets F., Schelstraete P., Haerynck F., Stove V., Van Simaeij L., Vaneecoutte M., Verstraete AG.**
Development and validation of an LC tandem MS assay for the quantification of beta-lactam antibiotics in the sputum of cystic fibrosis patients
Journal of Antimicrobial Chemotherapy 2018; 73: 95 - 101
- Fusco NM., Francisconi R., Meaney CJ., Duman D., Frederick CA., Prescott WA.**
Association of Vancomycin Trough Concentration with Response to Treatment for Acute Pulmonary Exacerbation of Cystic Fibrosis
Journal of the Pediatric Infectious Diseases Society 2017; 6: E103 - E108
- Ghorbani H., Memar MY., Sefidan FY., Yekani M., Ghotaslou R.**
In vitro synergy of antibiotic combinations against planktonic and biofilm *Pseudomonas aeruginosa*
GMS Hygiene and Infection Control 2017; 12: ArNo: Doc17
- Gupta PV., Nirwane AM., Belubbi T., Nagarsenker MS.**
Pulmonary delivery of synergistic combination of fluoroquinolone antibiotic complemented with proteolytic enzyme: A novel antimicrobial and antibiofilm strategy
Nanomedicine-nanotechnology Biology and Medicine 2017; 13: 2371 - 2384
- Haagensen J., Verotta D., Huang LS., Engel J., Spormann AM., Yang K.**
Spatiotemporal pharmacodynamics of meropenem- and tobramycin-treated *Pseudomonas aeruginosa* biofilms
Journal of Antimicrobial Chemotherapy 2017; 72: 3357 - 3365
- Habash MB., Goodyear MC., Park AJ., Surette MD., Vis EC., Harris RJ., Khursigara CM.**
Potentiation of Tobramycin by Silver Nanoparticles against *Pseudomonas aeruginosa* Biofilms
Antimicrobial Agents and Chemotherapy 2017; 61: 11:e00415-17
- Hoo ZH., Wildman MJ., Curley R., Walters SJ., Campbell MJ.**
Rescue therapy within the UK Cystic Fibrosis Registry: An exploration of predictors of intravenous antibiotic use amongst adults with CF
Respirology 2018; 23: 190 - 197
- Hurley MN., Fogarty A., McKeever TM., Goss CH., Rosenfeld M., Smyth AR.**
Early Respiratory Bacterial Detection and Antistaphylococcal Antibiotic Prophylaxis in Young Children with Cystic Fibrosis
Annals of the American Thoracic Society 2018; 15: 42 - 48
- Kerkhoff AD., Patrick L., Cornett P., Kleinhenz ME., Brondfield S.**
Severe piperacillin-tazobactam-induced hemolysis in a cystic fibrosis patient
Clinical Case Reports 2017; 5: 2059 - 2061
- Koeva M., Gutu AD., Hebert W., Wager JD., Yonker LM., O'Toole GA., Ausubel FM., Moskowitz SM., Joseph-McCarthy D.**
An Antipersister Strategy for Treatment of Chronic *Pseudomonas Aeruginosa* Infections
Antimicrobial Agents and Chemotherapy 2017; 61: 12:e00987-17
- Koo H., Allan RN., Howlin RP., Stoodley P., Hall-Stoodley L.**
Targeting microbial biofilms: current and prospective therapeutic strategies
Nature Reviews Microbiology 2017; 15: 740 - 755
- Le J., Bradley JS., Hingtgen S., Skochko S., Black N., Jones RN., Lim M., Capparelli EV.**
Pharmacokinetics of single-dose ceftaroline fosamil in children with cystic fibrosis
Pediatric Pulmonology 2017; 52: 1424 - 1434
- Li YT., Huang JR., Li LJ., Liu LS.**
Synergistic Activity of Berberine with Azithromycin against *Pseudomonas Aeruginosa* Isolated from Patients with Cystic Fibrosis of Lung In Vitro and In Vivo
Cellular Physiology and Biochemistry 2017; 42: 1657 - 1669
- Lopez-Causape C., de Dios-Caballero J., Cobo M., Escribano A., Asensio O., Oliver A., del Campo R., Canton R.**
Antibiotic resistance and population structure of cystic fibrosis *Pseudomonas aeruginosa* isolates from a Spanish multi-centre study
International Journal of Antimicrobial Agents 2017; 50: 334 - 341
- Lorenz J., Unnewehr M., Schaaf B., Gatermann S.**
Criteria for treating MRSA in sputum?
Internist 2017; 58: 1127 - 1132
- Maisetta G., Grassi L., Esin S., Serra I., Scordiapiino MA., Rinaldi AC., Batoni G.**
The Semi-Synthetic Peptide Lin-SB056-1 in Combination with EDTA Exerts Strong Antimicrobial and Antibiofilm Activity against *Pseudomonas aeruginosa* in Conditions Mimicking Cystic Fibrosis Sputum
International Journal of Molecular Sciences 2017; 18: 9:1994
- McKinzie CJ., Esther CR., Vece TJ.**
Continuous vancomycin in a pediatric cystic fibrosis patient
Pediatric Pulmonology 2018; 53: E4 - E5
- Mikalauskas A., Parkins MD., Poole K.**
Rifampicin potentiation of aminoglycoside activity against cystic fibrosis isolates of *Pseudomonas aeruginosa*
Journal of Antimicrobial Chemotherapy 2017; 72: 3349 - 3352

Morgan WJ., Wagener JS., Pasta DJ., Millar SJ., VanDevanter DR., Konstan MW.

Relationship of Antibiotic Treatment to Recovery after Acute FEV1 Decline in Children with Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 937 - 942

Perisson C., Destruys L., Grenet D., Bassinet L., Derelle J., Sermet-Gaudelus I., Thumerelle C., Prevotat A., Rosner V., Clement A., Corvol H.

Omalizumab treatment for allergic bronchopulmonary aspergillosis in young patients with cystic fibrosis
Respiratory Medicine 2017; 133: 12 - 15

Porsio B., Cusimano MG., Schillaci D., Craparo EF., Giammona G., Cavallaro G.

Nano into Micro Formulations of Tobramycin for the Treatment of Pseudomonas Aeruginosa Infections in Cystic Fibrosis
Biomacromolecules 2017; 18: 3924 - 3935

Pryjma M., Burian J., Kuchinski K., Thompson CJ.

Antagonism between Front-Line Antibiotics Clarithromycin and Amikacin in the Treatment of Mycobacterium abscessus Infections Is Mediated by the whiB7 Gene
Antimicrobial Agents and Chemotherapy 2017; 61: 11:e01353-17

Rac H., Stover KR., Wagner JL., King ST., Warnock HD., Barber KE.

Time-Kill Analysis of Ceftolozane/Tazobactam Efficacy Against Mucoid Pseudomonas Aeruginosa Strains from Cystic Fibrosis Patients
Infectious Diseases and Therapy 2017; 6: 507 - 513

Rouse C., Mistry P., Rayner O., Nickless J., Wan M., Southern KW., Batchelor HK.

A mixed methods study of the administration of flucloxacillin oral liquid; identifying strategies to overcome administration issues of medicines with poor palatability
International Journal of Pharmacy Practice 2017; 25: 326 - 334

Sanders DB., Solomon GM., Beckett VV., West NE., Danies CL., Heitshe SL., VanDevanter DR., Spahr JE., Gibson RL., Nick JA., Marshall BC., Flume PA., Goss CH.

Standardized Treatment of Pulmonary Exacerbations (STOP) study: Observations at the initiation of intravenous antibiotics for cystic fibrosis pulmonary exacerbations
Journal of Cystic Fibrosis 2017; 16: 592 - 599

Schniederjans M., Koska M., Haussler S.

Transcriptional and Mutational Profiling of an Aminoglycoside-Resistant Pseudomonas aeruginosa Small-Colony Variant
Antimicrobial Agents and Chemotherapy 2017; 61: 11:e01178-17

Shrestha CL., Assani KD., Rinehardt H., Albastroiu F., Zhang SZ., Shell R., Amer AO., Schlesinger LS., Kopp BT.

Cysteamine-mediated clearance of antibiotic-resistant pathogens in human cystic fibrosis macrophages
PLoS One 2017; 12: 10:e0186169

Singh M., Yau YCW., Wang S., Waters V., Kumar A.

MexXY efflux pump overexpression and aminoglycoside resistance in cystic fibrosis isolates of Pseudomonas aeruginosa from chronic infections

Canadian Journal of Microbiology 2017; 63: 929 - 938

Wang XT., Koehne-Voss S., Anumolu SS., Yu J.

Population Pharmacokinetics of Tobramycin Inhalation Solution in Pediatric Patients with Cystic Fibrosis
Journal of Pharmaceutical Sciences 2017; 106: 3402 - 3409

Waters V., Ratjen F.

Standard versus biofilm antimicrobial susceptibility testing to guide antibiotic therapy in cystic fibrosis
Cochrane Database of Systematic Reviews 2017; : 10:CD009528

Cardiology

Knight-Perry J., Branchford BR., Thornhill D., Martiniano SL., Sagel SD., Wang M.

Venous thromboembolism in children with cystic fibrosis: Retrospective incidence and intrapopulation risk factors
Thrombosis Research 2017; 158: 161 - 166

Urquhart DS., Vendrusculo FM.

Clinical interpretation of cardiopulmonary exercise testing in cystic fibrosis and implications for exercise counselling
Paediatric Respiratory Reviews 2017; 24: 72 - 78

Cell Biology

Garratt LW., Kicic A., Robertson C., Ranganathan S., Sly PD., Stick SM.

The AREST CF experience in biobanking - More than just tissues, tubes and time
Journal of Cystic Fibrosis 2017; 16: 622 - 627

Luft FC.

Cystic fibrosis: the conductance regulator, ceramides, and possible treatments
Journal of Molecular Medicine-jmm 2017; 95: 1017 - 1019

Merkert S., Bednarski C., Gohring G., Cathomen T., Martin U.

Generation of a gene-corrected isogenic control iPSC line from cystic fibrosis patient-specific iPSCs homozygous for p.Phe508del mutation mediated by TALENs and ssODN
Stem Cell Research 2017; 23: 95 - 97

Pierdomenico AM., Patruno S., Codagnone M., Simiele F., Mari VC., Plebani R., Recchiuti A., Romano M.

microRNA-181b is increased in cystic fibrosis cells and impairs lipoxin A(4) receptor-dependent mechanisms of inflammation resolution and antimicrobial defense
Scientific Reports 2017; 7: ArNo: 13519

Plebani R., Tripaldi R., Lanuti P., Recchiuti A., Patruno S., Di Silvestre S., Simeone P., Anile M., Venuta F., Prioletta M., Mucilli F., Del Porto P., Marchisio M., Pandolfi A., Romano M.

Establishment and long-term culture of human cystic fibrosis endothelial cells
Laboratory Investigation 2017; 97: 1375 - 1384

Puglia M., Landi C., Gagliardi A., Breslin L., Armini A., Brunetti J., Pini A., Bianchi L., Bini L.

The proteome speciation of an immortalized cystic fibrosis cell line: New perspectives on the pathophysiology of the disease
Journal of Proteomics 2018; 170: 28 - 42

Requena S., Ponomarchuk O., Castillo M., Rebik J., Brochiero E., Borejdo J., Gryczynski I., Dzyuba SV., Gryczynski Z., Grygorczyk R., Fudala R.

Imaging viscosity of intragranular mucin matrix in cystic fibrosis cells
Scientific Reports 2017; 7: ArNo: 16761

Schiumarini D., Loberto N., Mancini G., Bassi R., Giussani P., Chiricozzi E., Samarani M., Munari S., Tamanini A., Cabrini G., Lippi G., Dechechchi MC., Sonnino S., Aureli M.

Evidence for the Involvement of Lipid Rafts and Plasma Membrane Sphingolipid Hydrolases in Pseudomonas Aeruginosa Infection of Cystic Fibrosis Bronchial Epithelial Cells
Mediators of Inflammation 2017; : ArNo: 1730245

Schogler A., Blank F., Brugger M., Beyeler S., Tschanz SA., Regamey N., Casaulta C., Geiser T., Alves MP.

Characterization of pediatric cystic fibrosis airway epithelial cell cultures at the air-liquid interface obtained by non-invasive nasal cytology brush sampling
Respiratory Research 2017; 18: ArNo: 215

Totani L., Plebani R., Piccoli A., Di Silvestre S., Lanuti P., Recchiuti A., Cianci E., Dell'Elba G., Sacchetti S., Patruno S., Guarneri S., Mariggio MA., Mari VC., Anile M., Venuta F., Del Porto P., Moretti P., Prioletta M., et al
Mechanisms of endothelial cell dysfunction in cystic fibrosis
Biochimica et Biophysica Acta-Molecular Basis of Disease 2017; 1863: 3243 - 3253

CFTR

Agrawal PB., Wang RB., Li HMLS., Schmitz-Abe K., Simone-Roach C., Chen JX., Shi JH., Louie T., Sheng S., Towne MC., Brinson CF., Matthay MA., Kim CF., Bamshad M., Emond MJ., Gerard NP., Kleyman TR., Gerard C.
The Epithelial Sodium Channel Is a Modifier of the Long-Term Nonprogressive Phenotype Associated with F508del CFTR Mutations
American Journal of Respiratory Cell and Molecular Biology 2017; 57: 711 - 720

Ahmadi S., Bozoky Z., Di Paola M., Xia S., Li CH., Wong AP., Wellhauser L., Molinski SV., Ip W., Ouyang H., Avolio J., Forman-Kay JD., Ratjen F., Hirota JA., Rommens J., Rossant J., Gonska T., Moraes TJ., Bear CE.
Phenotypic profiling of CFTR modulators in patient-derived respiratory epithelia
NPJ Genomic Medicine 2017; 2: ArNo: 12

Arora K., Huang Y., Mun K., Yarlagadda S., Sundaram N., Kessler MM., Hannig G., Kurtz CB., Silos-Santiago I., Helmuth M., Palermo JJ., Clancy JP., Steinbrecher KA., Naren AP.
Guanylate cyclase 2C agonism corrects CFTR mutants
JCI Insight 2017; 2: 19:e93686

Avramescu RG., Kai Y., Xu HJ., Bidaud-Meynard A., Schnur A., Frenkiel S., Matouk E., Veit G., Lukacs GL.
Mutation-specific downregulation of CFTR2 variants by gating potentiaters
Human Molecular Genetics 2017; 26: 4873 - 4885

Bartoszewska S., Kamysz W., Jakiela B., Sanak M., Kroliczewski J., Bebok Z., Bartoszewski R., Collawn JF.
miR-200b downregulates CFTR during hypoxia in human lung epithelial cells
Cellular & Molecular Biology Letters 2017; 22: ArNo: 23

Benedetto R., Ousingsawat J., Wanitchakool P., Zhang Y., Holtzman MJ., Amaral M., Rock JR., Schreiber R., Kunzelmann K.
Epithelial Chloride Transport by CFTR Requires TMEM16A
Scientific Reports 2017; 7: ArNo: 12397

Callebaut I., Hoffmann B., Moron JP.
The implications of CFTR structural studies for cystic fibrosis drug development
Current Opinion in Pharmacology 2017; 34: 112 - 118

Chen H., Chan HC.
Amplification of FSH signalling by CFTR and nuclear soluble adenylyl cyclase in the ovary
Clinical and Experimental Pharmacology and Physiology 2017; 44:

DiFranco KM., Mulligan JK., Sumal AS., Diamond G.
Induction of CFTR gene expression by 1,25(OH)(2) vitamin D-3, 25OH vitamin D-3, and vitamin D-3 in cultured human airway epithelial cells and in mouse airways
Journal of Steroid Biochemistry and Molecular Biology 2017; 173: 323 - 332

Farhat R., El-Seedy A., Norez C., Talbot H., Pasquet MC., Adolphe C., Kitzis A., Ladeveze V.
Complexity of phenotypes induced by p.Asn1303Lys-CFTR correlates with difficulty to rescue and activate this protein
Cellular and Molecular Biology 2017; 63: 106 - 110

Farhat R., El-Seedy A., Sari AIP., Norez C., Pasquet MC., Becq F., Kitzis A., Ladeveze V.
In cellulo analyses of the p.Val322Ala mutation on the CFTR protein conformation and activity
Comptes Rendus Biologies 2017; 340: 367 - 371

Furgeri DT., Marson FAL., Correia CAA., Ribeiro JD., Bertuzzo CS.
Cystic fibrosis transmembrane regulator haplotypes in households of patients with cystic fibrosis
Gene 2018; 641: 137 - 143

Guimbellot JS., Leach JM., Chaudhry IG., Quinney NL., Boyles SE., Chua M., Aban I., Jaspers I., Gentzsch M.
Nasospheroids permit measurements of CFTR-dependent fluid transport
JCI Insight 2017; 2: 22:e95734

Hanrahan JW., Matthes E., Carlile G., Thomas DY.
Corrector combination therapies for F508del-CFTR
Current Opinion in Pharmacology 2017; 34: 105 - 111

Huang WQ., Jin AH., Zhang JT., Wang CQ., Tsang LL., Cai ZM., Zhou XP., Chen H., Chan HC.
Upregulation of CFTR in patients with endometriosis and its involvement in NF kappa B-uPAR dependent cell migration
Oncotarget 2017; 8: 66951 - 66959

Jih KY., Lin WY., Sohma Y., Hwang TC.
CFTR potentiaters: from bench to bedside
Current Opinion in Pharmacology 2017; 34: 98 - 104

Kido J., Shimohata T., Amano S., Hatayama S., Nguyen AQ., Sato Y., Kanda Y., Tentaku A., Fukushima S., Nakahashi M., Uebano T., Mawatari K., Takahashi A.
Cystic Fibrosis Transmembrane Conductance Regulator Reduces Microtubule-Dependent Campylobacter jejuni Invasion
Infection and Immunity 2017; 85: 10:e00311-17

Krishnan V., Maddox JW., Rodriguez T., Gleason E.
A role for the cystic fibrosis transmembrane conductance regulator in the nitric oxide-dependent release of Cl- from acidic organelles in amacrine cells
Journal of Neurophysiology 2017; 118: 2842 - 2852

Li HY., Salomon JJ., Sheppard DN., Mall MA., Galietta LJ.
Bypassing CFTR dysfunction in cystic fibrosis with alternative pathways for anion transport
Current Opinion in Pharmacology 2017; 34: 91 - 97

Li XP., Buonfiglio LGV., Adam RJ., Stoltz DA., Zabner J., Comellas AP.
Cystic Fibrosis Transmembrane Conductance Regulator Potentiation as a Therapeutic Strategy for Pulmonary Edema: A Proof-of-Concept Study in Pigs
Critical Care Medicine 2017; 45: e1240 - e1246

Li ZZ., Shen Z., Xue HP., Cheng S., Ji Q., Liu YT., Yang XJ.
CFTR protects against vascular inflammation and atherogenesis in apolipoprotein E-deficient mice
Bioscience Reports 2017; 37: ArNo: BSR20170

Lim SH., Legere EA., Snider J., Stagljar I.
Recent Progress in CFTR Interactome Mapping and Its Importance for Cystic Fibrosis
Frontiers in Pharmacology 2018; 8: ArNo: 997

Marengo B., Speciale A., Senatore L., Garibaldi S., Musumeci F., Nieddu E., Pollaro B., Pronzato MA., Schenone S., Mazzei M., Domenicotti C.
Matrine in association with FD-2 stimulates F508del-cystic fibrosis transmembrane conductance regulator activity in the presence of corrector VX809
Molecular Medicine Reports 2017; 16: 8849 - 8853

Mijnders M., Kleizen B., Braakman I.
Correcting CFTR folding defects by small-molecule correctors to cure cystic fibrosis
Current Opinion in Pharmacology 2017; 34: 83 - 90

- Oliver KE., Han ST., Sorscher EJ., Cutting GR.**
Transformative therapies for rare CFTR missense alleles
Current Opinion in Pharmacology 2017; 34: 76 - 82
- Oren YS., Pranke IM., Kerem B., Sermet-Gaudelus I.**
The suppression of premature termination codons and the repair of splicing mutations in CFTR
Current Opinion in Pharmacology 2017; 34: 125 - 131
- Prufert F., Bonengel S., Kollner S., Griesser J., Wilcox MD., Chater PI., Pearson JP., Bernkop-Schnurch A.**
zeta potential changing nanoparticles as cystic fibrosis transmembrane conductance regulator gene delivery system: an *in vitro* evaluation
Nanomedicine 2017; 12: 2713 - 2724
- Raju SV., Rasmussen L., Sloane PA., Tang LP., Libby EF., Rowe SM.**
Roflumilast reverses CFTR-mediated ion transport dysfunction in cigarette smoke-exposed mice
Respiratory Research 2017; 18: ArNo: 173
- Shi R., Xiao ZT., Zheng YJ., Zhang YL., Xu JW., Huang JH., Zhou WL., Li PB., Su WW.**
Naringenin Regulates CFTR Activation and Expression in Airway Epithelial Cells
Cellular Physiology and Biochemistry 2017; 44: 1146 - 1160
- Simhaev L., McCarty NA., Ford RC., Senderowitz H.**
Molecular Dynamics Flexible Fitting Simulations Identify New Models of the Closed State of the Cystic Fibrosis Transmembrane Conductance Regulator Protein
Journal of Chemical Information and Modeling 2017; 57: 1932 - 1946
- Smith E., Giuliano KA., Shumate J., Baillargeon P., McEwan B., Cullen MD., Miller JP., Drew L., Scampavia L., Spicer TP.**
A Homogeneous Cell-Based Halide-Sensitive Yellow Fluorescence Protein Assay to Identify Modulators of the Cystic Fibrosis Transmembrane Conductance Regulator Ion Channel
Assay and Drug Development Technologies 2017; :
- Solomon GM., Fu LW., Rowe SM., Collawn JF.**
The therapeutic potential of CFTR modulators for COPD and other airway diseases
Current Opinion in Pharmacology 2017; 34: 132 - 139
- Sorum B., Torocsik B., Csanady L.**
Asymmetry of movements in CFTR's two ATP sites during pore opening serves their distinct functions
eLife 2017; 6: ArNo: e29013
- Sun XS., Yi YL., Xie WL., Liang B., Winter MC., He N., Liu XM., Luo MH., Yang Y., Ode KL., Uc A., Norris AW., Engelhardt JF.**
CFTR Influences Beta Cell Function and Insulin Secretion Through Non-Cell Autonomous Exocrine-Derived Factors
Endocrinology 2017; 158: 3325 - 3338
- Tordai H., Leveles I., Hegedus T.**
Molecular dynamics of the cryo-EM CFTR structure
Biochemical and Biophysical Research Communications 2017; 491: 986 - 993
- Valdivieso AG., Mori C., Clauzure M., Massip-Copiz M., Santa-Coloma TA.**
CFTR modulates RPS27 gene expression using chloride anion as signaling effector
Archives of Biochemistry and Biophysics 2017; 633: 103 - 109
- Wedenöja S., Khamaysi A., Shimshilashvili L., Anbtawe-Jomaa S., Elomaa O., Toppari J., Hoglund P., Aittomaki K., Holmberg C., Hovatta O., Tapanainen JS., Ohana E., Kere J.**
A missense mutation in SLC26A3 is associated with human male subfertility and impaired activation of CFTR
Scientific Reports 2017; 7: ArNo: 14208
- Xavier BM., Hildebrandt E., Jiang F., Ding H., Kappes JC., Urbatsch IL.**
Substitution of Yor1p NBD1 residues improves the thermal stability of Human Cystic Fibrosis Transmembrane Conductance Regulator
Protein Engineering Design & Selection 2017; 30: 729 - 741
- Yeh HI., Sohma Y., Conrath K., Hwang TC.**
A common mechanism for CFTR potentiators
Journal of General Physiology 2017; 149: 1105 - 1118
- Zak SM., Clancy JP., Brewington JJ.**
CFTR functional assays in drug development
Expert Opinion on Orphan Drugs 2017; 5: 889 - 898
- Zeng M., Szymczak M., Ahuja M., Zheng CY., Yin HE., Swaim W., Chiorini JA., Bridges RJ., Muallem S.**
Restoration of CFTR Activity in Ducts Rescues Acinar Cell Function and Reduces Inflammation in Pancreatic and Salivary Glands of Mice
Gastroenterology 2017; 153: 1148 - 1159
- Zhang WQ., Zhang ZH., Zhang YH., Naren AP.**
CFTR-NHERF2-LPA(2) Complex in the Airway and Gut Epithelia
International Journal of Molecular Sciences 2017; 18: 9:1896
- Zhao D., Xu YZ., Li JT., Fu SE., Xiao FF., Song XW., Xie ZB., Jiang M., He Y., Liu CW., Wen QX., Yang XL.**
Association between F508 deletion in CFTR and chronic pancreatitis risk
Digestive and Liver Disease 2017; 49: 967 - 972
- Zhu Q., Li H., Liu Y., Jiang L.**
Knockdown of CFTR enhances sensitivity of prostate cancer cells to cisplatin via inhibition of autophagy
Neoplasma 2017; 64: 709 - 717

Clinical

- Briganti DF., D'Ovidio F.**
Long-term management of patients with end-stage lung diseases
Best Practice & Research-Clinical Anaesthesiology 2017; 31: 167 - 178
- Hobler MR., Engelberg RA., Curtis JR., Ramos KJ., Zander MI., Howard SS., Goss CH., Aitken ML.**
Exploring Opportunities for Primary Outpatient Palliative Care for Adults with Cystic Fibrosis: A Mixed-Methods Study of Patients' Needs
Journal of Palliative Medicine 2018; :
- Kerem E.**
Cystic fibrosis: Priorities and progress for future therapies
Paediatric Respiratory Reviews 2017; 24: 14 - 16
- Masson A., Kirszenbaum M., Sermet-Gaudelus I.**
Pain is an underestimated symptom in cystic fibrosis
Current Opinion in Pulmonary Medicine 2017; 23: 570 - 573
- Stanojevic S.**
Interpretation of Cystic Fibrosis Centre rankings: Meaningful comparisons or biased statistics?
Journal of Cystic Fibrosis 2017; 16: 534 - 535

Databases & Registries

- Claustres M., Theze C., des Georges M., Baux D., Girodon E., Bienvenu T., Audrezet MP., Dugueperoux I., Ferec C., Lalau G., Pagin A., Kitidis A., Thoreau V., Gaston V., Bieth E., Malinge MC., Reboul MP., Fergelot P., Lemonnier L., M**
CFTR-France, a national relational patient database for sharing genetic and phenotypic data associated with rare CFTR variants
Human Mutation 2017; 38: 1297 - 1315

- Fink AK., Loeffler DR., Marshall BC., Goss CH., Morgan WJ.**
Data that empower: The success and promise of CF patient registries
Pediatric Pulmonology 2017; 52:
- Jackson AD., Jackson AL., Fletcher G., Doyle G., Harrington M., Zhou SJ., Cullinane F., Gallagher C., McKone E.** Estimating Direct Cost of Cystic Fibrosis Care Using Irish Registry Healthcare Resource Utilisation Data, 2008-2012
Pharmacoconomics 2017; 35: 1087 - 1101
- Nightingale JA., Osmond C.**
Does current reporting of lung function by the UK cystic fibrosis registry allow a fair comparison of adult centres?
Journal of Cystic Fibrosis 2017; 16: 585 – 591

Diabetes

- Bogdani M., Blackman SM., Ridaura C., Belloc JP., Powers AC., Aguilar-Bryan L.**
Structural abnormalities in islets from very young children with cystic fibrosis may contribute to cystic fibrosis-related diabetes
Scientific Reports 2017; 7: ArNo: 17231
- Fattorusso V., Casale A., Raia V., Mozzillo E., Franzese A.**
Long-Term Follow-Up in a Girl with Cystic Fibrosis and Diabetes Since the First Year of Life
Diabetes Therapy 2017; 8: 1187 - 1190
- Haliloglu B., Gokdemir Y., Atay Z., Abali S., Guran T., Karakoc F., Ersu R., Karadag B., Turan S., Bereket A.**
Hypoglycemia is common in children with cystic fibrosis and seen predominantly in females
Pediatric Diabetes 2017; 18: 607 - 613
- Inman TB., Lim M., Proudfoot JA., Demeterco-Berggren C.**
Continuous glucose monitoring in a cystic fibrosis patient to predict pulmonary exacerbation?
Journal of Cystic Fibrosis 2017; 16: 628 - 630
- Leon MC., Gasso LB., Moreno-Galdo A., Martorrell AC., Tizzano SG., Fernandez DY., Lezcano AC.**
Oral glucose tolerance test and continuous glucose monitoring to assess diabetes development in cystic fibrosis patients
Endocrinologia Diabetes Y Nutricion 2018; 65: 45 - 51
- Moheet A., Moran A.**
CF-related diabetes: Containing the metabolic miscreant of cystic fibrosis
Pediatric Pulmonology 2017; 52:
- Mosnier-Pudar H.**
Cystic Fibrosis Related Diabetes (CFRD)
Correspondances en Metabolismes Hormones Diabetes et Nutrition 2017; 21: 237 - 240
- Reynaud Q., Poupon-Bourdy S., Rabilloud M., Al Mufti L., Jablonski CR., Lemonnier L., Nove-Josserand R., Touzet S., Durieu I.**
Pregnancy outcome in women with cystic fibrosis-related diabetes
Acta Obstetricia Et Gynecologica Scandinavica 2017; 96: 1223 - 1227
- Satin LS., Parekh VS.**
CFTR: Ferreting Out Its Role in Cystic Fibrosis-Related Diabetes
Endocrinology 2017; 158: 3319 – 3321

Diagnosis

- Basaran AE., Karatas-Torun N., Maslak IC., Bingol A., Alper OM.**
Normal sweat chloride test does not rule out cystic fibrosis
Turkish Journal of Pediatrics 2017; 59: 68 - 70

- Ciastkowska M., Ciastkowski M., Kalicki B.**
A new category of patients in the diagnostic process of cystic fibrosis
Pediatria I Medycyna Rodzinna-paediatrics and Family Medicine 2017; 13: 163 - 169
- Faria AG., Marson FAL., Gomez CCS., Servidoni MD., Ribeiro AF., Ribeiro JD.**
Thirty Years of Sweat Chloride Testing at One Referral Center
Frontiers in Pediatrics 2017; 5: ArNo: 222
- Gimpel C., Avni FE., Bergmann C., Cetiner M., Habbig S., Haffner D., Konig J., Konrad M., Liebau MC., Pape L., Rellensmann G., Titieni A., von Kaisenberg C., Weber S., Winyard PJD., Schaefer F.**
Perinatal Diagnosis, Management, and Follow-up of Cystic Renal Diseases A Clinical Practice Recommendation With Systematic Literature Reviews
JAMA Pediatrics 2018; 172: 74 - 86
- Salvatore M., Floridia G., Amato A., Censi F., de Stefano MC., Ferrari G., Tosto F., Taruscio D.**
Italian external quality assessment program for cystic fibrosis sweat chloride test: a 2015 and 2016 results comparison
Annali Dell Istituto Superiore Di Sanita 2017; 53: 305 - 313
- Talwar H., Hanoudi SN., Geamanu A., Kissner D., Draghici S., Samavati L.**
Detection of Cystic Fibrosis Serological Biomarkers Using a T7 Phage Display Library
Scientific Reports 2017; 7: ArNo: 17745

Endocrinology

- Cartault A., Tournier A., Ernoult P., Pienkowski C.**
Endocrine pathologies related to cystic fibrosis
Correspondances En Metabolismes Hormones Diabetes Et Nutrition 2017; 21: 241 – 245

Epidemiology

- Nunes LM., Ribeiro R., Niewiadonski VDT., Sabino E., Yamamoto GL., Bertola DR., Gaburo N., da Silva LVRF.**
A new insight into CFTR allele frequency in Brazil through next generation sequencing
Pediatric Pulmonology 2017; 52: 1300 - 1305
- Ortiz SC., Aguirre SJ., Flores S., Maldonado C., Mejia J., Salinas L.**
Spectrum of CFTR gene mutations in Ecuadorian cystic fibrosis patients: the second report of the p.H609R mutation
Molecular Genetics & Genomic Medicine 2017; 5: 751 - 757
- Poulimeneas D., Petrocheilou A., Grammatikopoulou MG., Kaditis AG., Loukou I., Doudounakis SE., Laggas D., Vassilakou T.**
High attainment of optimal nutritional and growth status observed among Greek pediatric cystic fibrosis patients: results from the GreeCF study
Journal of Pediatric Endocrinology & Metabolism 2017; 30: 1169 - 1176
- Stewart C., Pepper MS.**
Cystic Fibrosis in the African Diaspora
Annals of the American Thoracic Society 2017; 14: 1 – 7

Exercise

- Avramidou V., Hatziagorou E., Kampouras A., Hebestreit H., Kourouki E., Kirvassilis F., Tsanakas J.**
Lung clearance index (LCI) as a predictor of exercise limitation among CF patients
Pediatric Pulmonology 2018; 53: 81 - 87

Chelabi R., Soumagne T., Guillien A., Puyraveau M., Degano B.

In cystic fibrosis, lung clearance index is sensitive to detecting abnormalities appearing at exercise in children with normal spirometry
Respiratory Physiology & Neurobiology 2018; 247: 9 - 11

Vandekerckhove K., Keyzer M., Cornette J., Coomans I., Pyl F., De Baets F., Schelstraete P., Haerynck F., De Wolf D., Van Daele S., Boone J.

Exercise performance and quality of life in children with cystic fibrosis and mildly impaired lung function: relation with antibiotic treatments and hospitalization
European Journal of Pediatrics 2017; 176: 1689 – 1696

Gastroenterology

Ellemunter H., Engelhardt A., Schuller K., Steinkamp G.
Fecal Calprotectin in Cystic Fibrosis and Its Relation to Disease Parameters: A Longitudinal Analysis for 12 Years
Journal of Pediatric Gastroenterology and Nutrition 2017; 65: 438 - 442

Garg M., Leach ST., Coffey MJ., Katz T., Strachan R., Pang T., Needham B., Lui K., Ali F., Day AS., Appleton L., Moeeni V., Jaffe A., Ooi CY.

Age-dependent variation of fecal calprotectin in cystic fibrosis and healthy children
Journal of Cystic Fibrosis 2017; 16: 631 - 636

Hollander FM., de Roos NM., van Meerkerk GB., van Berkhouft FT., Heijerman HGM., van de Graaf EA.
Body Weight and Body Mass Index in Patients with End-Stage Cystic Fibrosis Stabilize After the Start of Enteral Tube Feeding
Journal of the Academy of Nutrition and Dietetics 2017; 117: 1808 - 1815

Houwen RHJ., van der Woerd WL., Slae M., Wilschanski M.
Effects of new and emerging therapies on gastrointestinal outcomes in cystic fibrosis
Current Opinion in Pulmonary Medicine 2017; 23: 551 - 555

Marson FAL., Bertuzzo CS., de Araujo TK., Hortencio TDR., Ribeiro AF., Ribeiro JD.

Pancreatic Insufficiency in Cystic Fibrosis: Influence of Inflammatory Response Genes
Pancreas 2018; 47: 99 - 109

Pagliari D., Saviano A., Serricchio ML., Dal Lago AA., Brizi MG., Manfredi R., Costamagna G., Attili F.
The association of pancreatic cystosis and IPMN in cystic fibrosis: case report and literature review
European Review for Medical and Pharmacological Sciences 2017; 21: 5179 - 5184

Piasecki B., Stanislawska-Kubiak M., Strzelecki W., Mojs E.
Attention and memory impairments in pediatric patients with cystic fibrosis and inflammatory bowel disease in comparison to healthy controls
Journal of Investigative Medicine 2017; 65: 1062 - 1067

Putman MS., Haagensen A., Neuringer I., Sicilian L.
Celiac Disease in Patients with Cystic Fibrosis-Related Bone Disease
Case Reports in Endocrinology 2017; : ArNo: 2652403

Tabori H., Jaudszus A., Arnold C., Mentzel HJ., Lorenz M., Michl RK., Lehmann T., Renz DM., Mainz JG.
Relation of Ultrasound Findings and Abdominal Symptoms obtained with the CFAbd-Score in Cystic Fibrosis Patients
Scientific Reports 2017; 7: ArNo: 17465

Gene Therapy

Carlon MS., Vidovic D., Birket S.

Roadmap for an early gene therapy for cystic fibrosis airway disease
Prenatal Diagnosis 2017; 37: 1181 - 1190

Cmielewski P., Farrow N., Devereux S., Parsons D., Donnelley M.

Gene therapy for Cystic Fibrosis: Improved delivery techniques and conditioning with lysophosphatidylcholine enhance lentiviral gene transfer in mouse lung airways
Experimental Lung Research 2017; 43: 426 - 433

Falese L., Sandza K., Yates B., Triffault S., Gangar S., Long B., Tsuruda L., Carter B., Vettermann C., Zoog SJ., Fong S.
Strategy to detect pre-existing immunity to AAV gene therapy
Gene Therapy 2017; 24: 768 - 778

Hart SL., Harrison PT.

Genetic therapies for cystic fibrosis lung disease
Current Opinion in Pharmacology 2017; 34: 119 - 124

Roesch EA., Drumm ML.

Powerful tools for genetic modification: Advances in gene editing
Pediatric Pulmonology 2017; 52:

Sasaki S., Guo SL.

Nucleic Acid Therapies for Cystic Fibrosis
Nucleic Acid Therapeutics 2017; :

Genetics

Chami H., Abou Arbid S., Badra R., Farra C.

A novel cystic fibrosis gene mutation c.2490insT in a Palestinian patient: A case report and review of the literature
Annals of Thoracic Medicine 2017; 12: 290 - 293

de Souza DAS., Faucz FR., Pereira-Ferrari L., Sotomaior VS., Raskin S.

Congenital bilateral absence of the vas deferens as an atypical form of cystic fibrosis: reproductive implications and genetic counseling
Andrology 2018; 6: 127 - 135

Gerbrands LC., Haarman EG., Hankel MA., Finken MJJ.

Cystic fibrosis and Silver-Russell syndrome due to a partial maternal isodisomy of chromosome 7
Clinical Case Reports 2017; 5: 1697 - 1700

Pereira SVN., Ribeiro JD., Bertuzzo CS., Marson FAL.

Association of clinical severity of cystic fibrosis with variants in the SLC gene family (SLC6A14, SLC26A9, SLC11A1 and SLC9A3)
Gene 2017; 629: 117 - 126

Terlizzi V., Di Lullo AM., Comegna M., Centrone C., Pelo E., Castaldo G., Raia V., Braggion C.

S737F is a new CFTR mutation typical of patients originally from the Tuscany region in Italy
Italian Journal of Pediatrics 2018; 44:

Yao Y., Feng XL., Xu BP., Shen KL.

Pseudo-Bartter Syndrome in a Chinese Infant with Cystic Fibrosis Caused by c.532G > A Mutation in CFTR
Chinese Medical Journal 2017; 130: 2771 – 2772

Growth & Development

Gruet M., Troosters T., Verges S.

Peripheral muscle abnormalities in cystic fibrosis: Etiology, clinical implications and response to therapeutic interventions
Journal of Cystic Fibrosis 2017; 16: 538 - 552

- Gupta S., Kabra SK.**
Bone Mineral Density in Cystic Fibrosis: Few Concerns Reply
Indian Pediatrics 2017; 54: 973 - 974
- Kenis-Coskun O., Karadag-Saygi E., Bahar-Ozdemir Y., Gokdemir Y., Karadag B., Kayhan O.**
The involvement of musculoskeletal system and its influence on postural stability in children and young adults with cystic fibrosis
Italian Journal of Pediatrics 2017; 43: ArNo: 106
- Siddiqui SA.**
Bone Mineral Density in Cystic Fibrosis: Few Concerns
Indian Pediatrics 2017; 54: 973
- Smith N., Lim A., Yap M., King L., James S., Jones A., Ranganathan S., Simm P.**
Bone mineral density is related to lung function outcomes in young people with cystic fibrosis-A retrospective study
Pediatric Pulmonology 2017; 52: 1558 - 1564
- Tomlinson OW., Barker AR., Oades PJ., Williams CA.**
Scaling the Oxygen Uptake Efficiency Slope for Body Size in Cystic Fibrosis
Medicine and Science in Sports and Exercise 2017; 49: 1980 - 1986
- Giddings O., Esther CR.**
Mapping targetable inflammation and outcomes with cystic fibrosis biomarkers
Pediatric Pulmonology 2017; 52:
- Gothe F., Kappler M., Gries M.**
Increasing Total Serum IgE, Allergic Bronchopulmonary Aspergillosis, and Lung Function in Cystic Fibrosis
Journal of Allergy and Clinical Immunology-in Practice 2017; 5: 1591 - +
- Gurczynski SJ., Moore BB.**
IL-17 in the lung: the good, the bad, and the ugly
American Journal of Physiology-lung Cellular and Molecular Physiology 2018; 314: L6 - L16
- Khouri O., Barrios C., Ortega V., Atala A., Murphy SV.**
Immunomodulatory Cell Therapy to Target Cystic Fibrosis Inflammation
American Journal of Respiratory Cell and Molecular Biology 2018; 58: 12 - 20
- Krick S., Baumlin N., Aller SP., Aguiar C., Grabner A., Sailland J., Mendes E., Schmid A., Qi LX., David NV., Geraghty P., King G., Birket SE., Rowe SM., Faul C., Salathe M.**
Klotho Inhibits Interleukin-8 Secretion from Cystic Fibrosis Airway Epithelia
Scientific Reports 2017; 7: ArNo: 14388
- Laguna TA., Williams CB., Nunez MG., Welchlin-Bradford C., Moen CE., Reilly CS., Wendt CH.**
Biomarkers of inflammation in infants with cystic fibrosis
Respiratory Research 2018; 19: ArNo: 6
- Law SM., Gray RD.**
Neutrophil extracellular traps and the dysfunctional innate immune response of cystic fibrosis lung disease: a review
Journal of Inflammation-London 2017; 14: ArNo: 29
- Lucca F., Da Dalt L., Ros M., Gucciardi A., Pirillo P., Naturale M., Perilongo G., Giordano G., Baraldi E.**
Asymmetric dimethylarginine and related metabolites in exhaled breath condensate of children with cystic fibrosis
Clinical Respiratory Journal 2018; 12: 140 - 148
- Pang Z., Junkins RD., MacNeil AJ., McCormick C., Cheng ZY., Chen WM., Lin TJ.**
The calcineurin-NFAT axis contributes to host defense during *Pseudomonas aeruginosa* lung infection
Journal of Leukocyte Biology 2017; 102: 1461 - 1469
- Piasecki B., Stanislawska-Kubiak M., Strzelecki W., Mojs E.**
Attention and memory impairments in pediatric patients with cystic fibrosis and inflammatory bowel disease in comparison to healthy controls
Journal of Investigative Medicine 2017; 65: 1062 - 1067
- Polineni D., Dang H., Gallins PJ., Jones LC., Pace RG., Stonebraker JR., Commander LA., Krenicky JE., Zhou YH., Corvol H., Cutting GR., Drumm ML., Strug LJ., Boyle MP., Durie PR., Chmiel JF., Zou F., Wright FA., O'Neal WK., et al**
Airway Mucosal Host Defense Is Key to Genomic Regulation of Cystic Fibrosis Lung Disease Severity
American Journal of Respiratory and Critical Care Medicine 2018; 197: 79 - 93
- Saint-Criq V., Villeret B., Bastaert F., Kheir S., Hatton A., Cazes A., Xing Z., Sermet-Gaudelus I., Garcia-Verdugo I., Edelman A., Sallenave JM.**
Pseudomonas aeruginosa LasB protease impairs innate immunity in mice and humans by targeting a lung epithelial cystic fibrosis transmembrane regulator-IL-6-antimicrobial-repair pathway
Thorax 2018; 73: 49 - 61
- ## Immunology & Inflammation
- Abid S., Xie SK., Bose M., Shaul PW., Terada LS., Brody SL., Thomas PJ., Katzenellenbogen JA., Kim SH., Greenberg DE., Jain R.**
17 beta-Estradiol Dysregulates Innate Immune Responses to *Pseudomonas Aeruginosa* Respiratory Infection and Is Modulated by Estrogen Receptor Antagonism
Infection and Immunity 2017; 85: 10:e00422-17
- Becker KA., Li X., Seitz A., Steinmann J., Koch A., Schuchman E., Kamler M., Edwards MJ., Caldwell CC., Gulbins E.**
Neutrophils Kill Reactive Oxygen Species-Resistant *Pseudomonas aeruginosa* by Sphingosine
Cellular Physiology and Biochemistry 2017; 43: 1603 - 1616
- Bodas M., Mazur S., Min T., Vij N.**
Inhibition of histone-deacetylase activity rescues inflammatory cystic fibrosis lung disease by modulating innate and adaptive immune responses
Respiratory Research 2018; 19: ArNo: 2
- Boikos C., Joseph L., Scheifele D., Lands LC., De Serres G., Papenburg J., Winters N., Chilvers M., Quach C.**
Adverse events following live-attenuated intranasal influenza vaccination of children with cystic fibrosis: Results from two influenza
Vaccine 2017; 35: 5019 - 5026
- Clerc A., Reynaud Q., Durupt S., Chapuis-Cellier C., Nove-Josserand R., Durieu I., Lega JC.**
Elevated IgG4 serum levels in patients with cystic fibrosis
PLoS One 2017; 12: 9:e0181888
- Dourbes G., Berger P., Macey JRJ., Bui S., Delhaes L., Montaudon M., Corneloup O., Chateil JF., Marthan R., Fayon M., Laurent F.**
Allergic Bronchopulmonary Aspergillosis in Cystic Fibrosis: VR Imaging of Airway Vucus Contrasts as a Tool for Diagnosis
Radiology 2017; 285: 261 - 269
- Furlan LL., Ribeiro JD., Bertuzzo CS., Salomao JB., Souza DRS., Marson FAL.**
Variants in the interleukin 8 gene and the response to inhaled bronchodilators in cystic fibrosis
Jornal de Pediatria 2017; 93: 639 - 648

Sellers ZM., Illek B., Figueira MF., Hari G., Joo NS., Sibley E., Souza-Menezes J., Morales MM., Fischer H., Wine JJ.
Impaired PGE(2)-stimulated Cl- and HCO3- secretion contributes to cystic fibrosis airway disease
PLoS One 2017; 12: e0189894

Svedin E., Utorova R., Huhn MH., Larsson PG., Stone VM., Garimella M., Lind K., Hagglof T., Pincikova T., Laitinen OH., McInerney GM., Scholte B., Hjelte L., Karlsson MCI., Flodstrom-Tullberg M.
A Link Between a Common Mutation in CFTR and Impaired Innate and Adaptive Viral Defense
Journal of Infectious Diseases 2017; 216: 1308 - 1317

Tynan A., Mawhinney L., Armstrong ME., O'Reilly C., Kennedy S., Caraher E., Julicher K., O'Dwyer D., Maher L., Schaffer K., Fabre A., McKone EF., Leng L., Bucala R., Bernhagen J., Cooke G., Donnelly SC.
Macrophage migration inhibitory factor enhances *Pseudomonas aeruginosa* biofilm formation, potentially contributing to cystic fibrosis pathogenesis
FASEB Journal 2017; 31: 5102 - 5110

Uyan ZS., Unluguzel Ustun G., Haklar G., Cakir E., Oktem S., Ersu R., Karadag BT., Karakoc F., Dagli E.
Effect of inhaled steroids on clinical and inflammatory parameters in children with cystic fibrosis
Turkish Journal of Medical Sciences 2017; 47: 1432 - 1440

Vargas MH., Del-Razo-Rodriguez R., Lopez-Garcia A., Lezana-Fernandez JL., Chavez J., Furuya MEY., Marin-Santana JC.
Effect of oral glycine on the clinical, spirometric and inflammatory status in subjects with cystic fibrosis: a pilot randomized trial
BMC Pulmonary Medicine 2017; 17: ArNo: 206

Liver Disease

Cheng K., Ashby D., Smyth RL.
Ursodeoxycholic acid for cystic fibrosis-related liver disease (Review)
Cochrane Database of Systematic Reviews 2017; : 9:CD000222

Drzymala-Czyz S., Szczepanik M., Krzyzanowska P., Dusz-Zuchowska M., Pogorzelski A., Sapiejka E., Juszcza P., Lisowska A., Koletzko B., Walkowiak J.
Serum Phospholipid Fatty Acid Composition in Cystic Fibrosis Patients with and without Liver Cirrhosis
Annals of Nutrition and Metabolism 2017; 71: 91 - 98

Feige J., Mainz JG., Tabori H., Renz DM., Stenzel M., Mentzel HJ.
ARFI elastography of cystic fibrosis-related liver disease (CFLD)
Monatsschrift Kinderheilkunde 2017; 165: 888 - 894

Pozniak KN., Pearen MA., Pereira TN., Kramer CSM., Kalita-De Croft P., Nawaratna SK., Fernandez-Rojo MA., Gobert GN., Tirnitz-Parker JEE., Olynyk JK., Shepherd RW., Lewindon PJ., Runnel GA.
Taurocholate Induces Biliary Differentiation of Liver Progenitor Cells Causing Hepatic Stellate Cell Chemotaxis in the Ductular Reaction Role in Pediatric Cystic Fibrosis Liver Disease
American Journal of Pathology 2017; 187: 2744 - 2757

van de Peppel IP., Bertolini A., Jonker JW., Bodewes FAJA., Verkade HJ.
Diagnosis, follow-up and treatment of cystic fibrosis-related liver disease
Current Opinion in Pulmonary Medicine 2017; 23: 562 - 569

Meeting Abstracts

[Anonymous].

Abstracts from the 23rd Italian congress of Cystic Fibrosis and the 13th National congress of Cystic Fibrosis Italian Society Naples, Italy. 22-25 November 2017 Abstracts
Italian Journal of Pediatrics 2018; 44

Microbiology

Abdalla MY., Hoke T., Seravalli J., Switzer BL., Bavitz M., Fliege JD., Murphy PJ., Britigan BE.

Pseudomonas Quinolone Signal Induces Oxidative Stress and Inhibits Heme Oxygenase-1 Expression in Lung Epithelial Cells
Infection and Immunity 2017; 85: 9:e00176-17

Acosta N., Whelan FJ., Somayaji R., Poonja A., Surette MG., Rabin HR., Parkins MD.

The Evolving Cystic Fibrosis Microbiome: A Comparative Cohort Study Spanning 16 Years
Annals of the American Thoracic Society 2017; 14: 1288 - 1297

Aditi., Shariff M., Chhabra SK., Rahman MU.

Similar virulence properties of infection and colonization associated *Pseudomonas aeruginosa*
Journal of Medical Microbiology 2017; 66: 1489 - 1498

Amaya PP., Haim MS., Fernandez S., Di Gregorio S., Teper A., Vazquez M., Lubovich S., Galanternik L., Mollerach M.

Molecular Epidemiology of Methicillin-Resistant *Staphylococcus aureus* in Cystic Fibrosis Patients from Argentina
Microbial Drug Resistance 2017; :

Baker YR., Hodgkinson JT., Florea BI., Alza E., Galloway WRJD., Grimm L., Geddis SM., Overkleef HS., Welch M., Spring DR.

Identification of new quorum sensing autoinducer binding partners in *Pseudomonas aeruginosa* using photoaffinity probes
Chemical Science 2017; 8: 7403 - 7411

Barsky EE., Williams KA., Priebe GP., Sawicki GS.

Incident *Stenotrophomonas maltophilia* infection and lung function decline in cystic fibrosis
Pediatric Pulmonology 2017; 52: 1276 - 1282

Beaudoin T., Yau YCW., Stapleton PJ., Gong Y., Wang PW., Guttman DS., Waters V.

Staphylococcus aureus interaction with *Pseudomonas aeruginosa* biofilm enhances tobramycin resistance
NPJ Biofilms and Microbiomes 2017; 3: ArNo: UNSP 25

Boutin S., Depner M., Stahl M., Graeber SY., Dittrich SA., Legatzki A., von Mutius E., Mall M., Dalpke AH.

Comparison of Oropharyngeal Microbiota from Children with Asthma and Cystic Fibrosis
Mediators of Inflammation 2017; : ArNo: 5047403

Butt AT., Thomas MS.

Iron Acquisition Mechanisms and Their Role in the Virulence of *Burkholderia* Species
Frontiers in Cellular and Infection Microbiology 2017; 7: ArNo: 460

Caballero JD., Vida R., Cobo M., Maiz L., Suarez L., Galeano J., Baquero F., Canton R., del Campo R.

Individual Patterns of Complexity in Cystic Fibrosis Lung Microbiota, Including Predator Bacteria, over a 1-Year Period
mBio 2017; 8: 5:e00959-17

Capizzani CPD., Cacador NC., Torres LAGMM., Tonani L., Vandamme P., Darini ALD.

Clinical and microbiological profile of chronic *Burkholderia cepacia* complex infections in a cystic fibrosis reference hospital in Brazil
European Journal of Clinical Microbiology & Infectious Diseases 2017; 36: 2263 - 2271

- Carsin A., Romain T., Ranque S., Reynaud-Gaubert M., Dubus JC., Mege JL., Vitte J.**
Aspergillus fumigatus in cystic fibrosis: An update on immune interactions and molecular diagnostics in allergic bronchopulmonary aspergillosis
Allergy 2017; 72: 1632 - 1642
- Caskey S., Stirling J., Moore JE., Rendall JC.**
Occurrence of Pseudomonas aeruginosa in taps: implications for patients with cystic fibrosis
Journal of Hospital Infection 2018; 98: 64 - 65
- Cavalli Z., Reynaud Q., Bricca R., Nove-Josserand R., Durupt S., Reix P., Perceval M., de Montclos MP., Lina G., Durieu I.**
High incidence of non-tuberculous mycobacteria-positive cultures among adolescent with cystic fibrosis
Journal of Cystic Fibrosis 2017; 16: 579 - 584
- Cohen RWF., Folescu TW., Daltro P., Boechat MCB., Lima DF., Marques EA., Leao RS.**
Methicillin-resistant Staphylococcus aureus in cystic fibrosis patients: do we need to care? A cohort study
Sao Paulo Medical Journal 2017; 135: 420 - 427
- Dentini P., Marson FAL., Bonadia LC., Bertuzzo CS., Ribeiro AF., Levy CE., Ribeiro JD.**
Burkholderia cepacia complex in cystic fibrosis in a Brazilian reference center
Medical Microbiology and Immunology 2017; 206: 447 - 461
- Di Paola M., Park AJ., Ahmadi S., Roach EJ., Wu YS., Struderman K., Lam JS., Bear CE., Khursigara CM.**
SLC6A14 Is a Genetic Modifier of Cystic Fibrosis That Regulates Pseudomonas Aeruginosa Attachment to Human Bronchial Epithelial Cells
mBio 2017; 8: e02073-17
- Dittrich AM.**
Chronic Pseudomonas aeruginosa airway colonization in cystic fibrosis patients: Prevention concepts
Internist 2017; 58: 1133 - 1141
- Dupont C., Aujoulat F., Chiron R., Condom P., Jumas-Bilak E., Marchandin H.**
Highly Diversified Pandoraea pulmonicola Population during Chronic Colonization in Cystic Fibrosis
Frontiers in Microbiology 2017; 8: ArNo: 1892
- Fila L., Drevinek P.**
Burkholderia cepacia complex in cystic fibrosis in the post-epidemic period: multilocus sequence typing-based approach
Folia Microbiologica 2017; 62: 509 - 514
- Frayman KB., Armstrong DS., Carzino R., Ferkol TW., Grimwood K., Storch GA., Teo SM., Wylie KM., Ranganathan SC.**
The lower airway microbiota in early cystic fibrosis lung disease: a longitudinal analysis
Thorax 2017; 72: 1104 - 1112
- Frayman KB., Armstrong DS., Grimwood K., Ranganathan SC.**
The airway microbiota in early cystic fibrosis lung disease
Pediatric Pulmonology 2017; 52: 1384 - 1404
- Gao YT., Duan JJ., Geng X., Zhang ZQ., Zhang RQ., Li XX., Wang SY., Kang JB., Yin DH., Song Y.**
Deficiency of quorum sensing system inhibits the resistance selection of Pseudomonas aeruginosa to ciprofloxacin and levofloxacin in vitro
Journal of Global Antimicrobial Resistance 2017; 10: 113 - 119
- Green HD., Bright-Thomas R., Kenna DT., Turton JF., Woodford N., Jones AM.**
Ralstonia infection in cystic fibrosis
Epidemiology and Infection 2017; 145: 2864 - 2872
- Groitl B., Dahl JU., Schroeder JW., Jakob U.**
Pseudomonas aeruginosa defense systems against microbicidal oxidants
Molecular Microbiology 2017; 106: 335 - 350
- Hosseinkhan N., Mousavian Z., Masoudi-Nejad A.**
Comparison of gene co-expression networks in Pseudomonas aeruginosa and *Staphylococcus aureus* reveals conservation in some aspects of virulence
Gene 2018; 639: 1 - 10
- Hurley MN., Fogarty A., McKeever TM., Goss CH., Rosenfeld M., Smyth AR.**
Early Respiratory Bacterial Detection and Antistaphylococcal Antibiotic Prophylaxis in Young Children with Cystic Fibrosis
Annals of the American Thoracic Society 2018; 15: 42 - 48
- Hvorecny KL., Dolben E., Moreau-Marquis S., Hampton TH., Shabaneh TB., Flitter BA., Bahl CD., Bomberger JM., Levy BD., Stanton BA., Hogan DA., Madden DR.**
An epoxide hydrolase secreted by Pseudomonas aeruginosa decreases mucociliary transport and hinders bacterial clearance from the lung
American Journal of Physiology-lung Cellular and Molecular Physiology 2018; 314: L150 - L156
- Joest M., Sennekamp J.**
Allergic bronchopulmonary aspergillosis (ABPA) and other allergic bronchopulmonary mycoses
Allergologie 2017; 40: 522 - 534
- Johnson S., McNeal M., Mermis J., Polineni D., Burger S.**
Chasing Zero: Increasing Infection Control Compliance on an Inpatient Cystic Fibrosis Unit
Journal of Nursing Care Quality 2018; 33: 67 - 71
- Kamath KS., Krisp C., Chick J., Pascovici D., Gygi SP., Molloy MP.**
Pseudomonas aeruginosa Proteome under Hypoxic Stress Conditions Mimicking the Cystic Fibrosis Lung
Journal of Proteome Research 2017; 16: 3917 - 3928
- Korten I., Kieninger E., Klenja S., Mack I., Schlapfer N., Barbani MT., Regamey N., Kuehni CE., Hilty M., Frey U., Gorgievski M., Casaulta C., Latzin P.**
Respiratory viruses in healthy infants and infants with cystic fibrosis: a prospective cohort study
Thorax 2018; 73: 13 - 20
- Kovach K., Davis-Fields M., Irie Y., Jain K., Doorwar S., Vuong K., Dhamani N., Mohanty K., Touhami A., Gordon VD.**
Evolutionary adaptations of biofilms infecting cystic fibrosis lungs promote mechanical toughness by adjusting polysaccharide production
NPI Biofilms and Microbiomes 2017; 3: ArNo: UNSP 1
- Lauridsen RK., Sommer LM., Johansen HK., Rindzevicius T., Molin S., Jelsbak L., Engelsen SB., Boisen A.**
SERS detection of the biomarker hydrogen cyanide from Pseudomonas aeruginosa cultures isolated from cystic fibrosis patients
Scientific Reports 2017; 7: ArNo: 45264
- Li H., Li X., Song C., Zhang Y., Wang Z., Liu Z., Wei H., Yu J.**
Autoinducer-2 Facilitates Pseudomonas aeruginosa PAO1 Pathogenicity in Vitro and in Vivo
Frontiers in Microbiology 2017; 8: ArNo: 1944
- Londino JD., Lazrak A., Collawn JF., Bebok Z., Harrod KS., Matalon S.**
Influenza virus infection alters ion channel function of airway and alveolar cells: mechanisms and physiological sequelae
American Journal of Physiology-Lung Cellular and Molecular Physiology 2017; 313: L845 - L858

- Lopes SP., Azevedo NF., Pereir MO.**
Developing a model for cystic fibrosis sociomicrobiology based on antibiotic and environmental stress
International Journal of Medical Microbiology 2017; 307: 460 - 470
- Macin S., Akarca M., Sener B., Akyon Y.**
Comparison of virulence factors and antibiotic resistance of *Pseudomonas aeruginosa* strains isolated from patients with and without cystic fibrosis
Revista Romana de Medicina de Laborator 2017; 25: 327 - 334
- Maggini V., Pesavento G., Maida I., Lo Nostro A., Calonico C., Sassoli C., Perrin E., Fondi M., Mengoni A., Chiellini C., Vannacci A., Gallo E., Gori L., Bogani P., Bilia AR., Campana S., Ravenni N., Dolce D., Firenzuoli F., et al**
Exploring the Effect of the Composition of Three Different Oregano Essential Oils on the Growth of Multidrug-Resistant Cystic Fibrosis *Pseudomonas aeruginosa* Strains
Natural Product Communications 2017; 12: 1949 - 1952
- Maille E., Ruffin M., Adam D., Messaoud H., Lafayette SL., McKay G., Nguyen D., Brochiero E.**
Quorum Sensing Down-Regulation Counteracts the Negative Impact of *Pseudomonas aeruginosa* on CFTR Channel Expression, Function and Rescue in Human Airway Epithelial Cells
Frontiers in Cellular and Infection Microbiology 2017; 7: ArNo: 470
- Marmont LS., Whitfield GB., Rich JD., Yip P., Giesbrecht LB., Stremick CA., Whitney JC., Parsek MR., Harrison JJ., Howell PL.**
PelA and PelB proteins form a modification and secretion complex essential for Pel polysaccharide-dependent biofilm formation in *Pseudomonas aeruginosa*
Journal of Biological Chemistry 2017; 292: 19411 - 19422
- Martiniano SL., Davidson RM., Nick JA.**
Nontuberculous mycobacteria in cystic fibrosis: Updates and the path forward
Pediatric Pulmonology 2017; 52:
- Millar BC., McCaughan J., Rendall JC., Downey DG., Moore JE.**
Pseudomonas aeruginosa in cystic fibrosis patients with c.1652G > A (G551D)-CFTR treated with ivacaftor Changes in microbiological parameters
Journal of Clinical Pharmacy and Therapeutics 2018; 43: 92 - 100
- Mina S., Staerck C., Marot A., Godon C., Calenda A., Bouchara JP., Fleury MJJ.**
Scedosporium boydii CatA1 and SODC recombinant proteins, new tools for serodiagnosis of Scedosporium infection of patients with cystic fibrosis
Diagnostic Microbiology and Infectious Disease 2017; 89: 282 - 287
- Montero AV., de Vicente CM., Romero RG.**
Acute pancreatitis caused by *Campylobacter jejuni* in a child with cystic fibrosis
Medicina Clinica 2017; 149: 513 - 514
- Moreira AS., Mil-Homens D., Sousa SA., Coutinho CP., Pinto-de-Oliveira A., Ramosy CG., dos Santos SC., Fialho AM., Leitao JH., Sa-Correia I.**
Variation of *Burkholderia* cenocepacia virulence potential during cystic fibrosis chronic lung infection
Virulence 2017; 8: 782 - 796
- Muhlebach MS.**
Methicillin-resistant *Staphylococcus aureus* in cystic fibrosis: how should it be managed?
Current Opinion in Pulmonary Medicine 2017; 23: 544 - 550
- Nasir M., Bean HD., Smolinska A., Rees CA., Zemanick ET., Hill JE.**
Volatile molecules from bronchoalveolar lavage fluid can 'rule-in' *Pseudomonas aeruginosa* and 'rule-out' *Staphylococcus aureus* infections in cystic fibrosis patients
Scientific Reports 2018; 8: ArNo: 826
- Nunvar J., Capek V., Fiser K., Fila L., Drevinek P.**
What matters in chronic *Burkholderia* cenocepacia infection in cystic fibrosis: Insights from comparative genomics
Plos Pathogens 2017; 13: 12:e1006762
- Olszak T., Shneider MM., Latka A., Maciejewska B., Browning C., Sycheva LV., Cornelissen A., Danis-Wlodarczyk K., Senchenkova SN., Shashkov AS., Gula G., Arabski M., Wasik S., Miroshnikov KA., Lavigne R., Leiman PG., Knirel YA., et al**
The O-specific polysaccharide lyase from the phage LKA1 tailspike reduces *Pseudomonas* virulence
Scientific Reports 2017; 7: ArNo: 16302
- Radlinski L., Rowe SE., Kartchner LB., Maile R., Cairns BA., Vitko NP., Gode CJ., Lachiewicz AM., Wolfgang MC., Conlon BP.**
Pseudomonas aeruginosa exoproducts determine antibiotic efficacy against *Staphylococcus aureus*
Plos Biology 2017; 15: 11:e2003981
- Rado J., Kaszab E., Petrovics T., Paszti J., Kriszt B., Szoboszlay S.**
Characterization of environmental *Pseudomonas aeruginosa* using multilocus sequence typing scheme
Journal of Medical Microbiology 2017; 66: 1457 - 1466
- Reynaud Q., Dupont D., Nove-Josserand R., Durupt S., Persat F., Ader F., Grenet D., Durieu I.**
Rare and unusual presentation of Cladophialophora infection in a pulmonary transplant cystic fibrosis patient
Transplant Infectious Disease 2017; 19: 6:e12789
- Rogers GB., Bruce KD., Hoffman LR.**
How can the cystic fibrosis respiratory microbiome influence our clinical decision-making?
Current Opinion in Pulmonary Medicine 2017; 23: 536 - 543
- Silva IN., Ramires MJ., Azevedo LA., Guerreiro AR., Tavares AC., Becker JD., Moreira LM.**
Regulator LdhR and D-Lactate Dehydrogenase LdhA of *Burkholderia multivorans* Play Roles in Carbon Overflow and in Planktonic Cellular Aggregate Formation
Applied and Environmental Microbiology 2017; 83: 19:UNSP e01343
- Skolnik K., Nguyen A., Thornton CS., Waddell B., Williamson T., Rabin HR., Parkins MD.**
Group B streptococcus (GBS) is an important pathogen in human disease-but what about in cystic fibrosis?
BMC Infectious Diseases 2017; 17: ArNo: 660
- Smith AC., Rice A., Sutton B., Gabrilska R., Wessel AK., Whiteley M., Rumbaugh KP.**
Albumin Inhibits *Pseudomonas aeruginosa* Quorum Sensing and Alters Polymicrobial Interactions
Infection and Immunity 2017; 85: 9:e00116-17
- Somayaji R., Stanojevic S., Tullis DE., Stephenson AL., Ratjen F., Waters V.**
Clinical Outcomes Associated with *Achromobacter* Species Infection in Patients with Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 1412 - 1418
- Stefani S., Campana S., Cariani L., Carnovale V., Colombo C., Lleo MM., Iula VD., Minicucci L., Morelli P., Pizzamiglio G., Taccetti G.**
Relevance of multidrug-resistant *Pseudomonas aeruginosa* infections in cystic fibrosis
International Journal of Medical Microbiology 2017; 307: 353 - 362

Stietz MS., Lopez C., Osifo O., Tolmasky ME., Cardona ST.
Evaluation of the electron transfer flavoprotein as an antibacterial target in Burkholderia cenocepacia
Canadian Journal of Microbiology 2017; 63: 857 - 863

Tai AS., Sherrard LJ., Kidd TJ., Ramsay KA., Buckley C., Syrmis M., Grimwood K., Bell SC., Whiley DM.
Antibiotic perturbation of mixed-strain Pseudomonas aeruginosa infection in patients with cystic fibrosis
BMC Pulmonary Medicine 2017; 17: ArNo: 138

Trovalo A., Baldan R., Costa D., Simonetti TM., Cirillo DM., Tortoli E.
Molecular Typing of Mycobacterium Abscessus Isolated from Cystic Fibrosis Patients
International Journal of Mycobacteriology 2017; 6: 138 - 141

Vernocchi P., Del Chierico F., Quagliariello A., Ercolini D., Lucidi V., Putignani L.
A Metagenomic and in Silico Functional Prediction of Gut Microbiota Profiles May Concur in Discovering New Cystic Fibrosis Patient- Targeted Probiotics
Nutrients 2017; 9: 12:1342

Wijers CDM., Chmiel JF., Gaston BM.
Bacterial infections in patients with primary ciliary dyskinesia: Comparison with cystic fibrosis
Chronic Respiratory Disease 2017; 14: 392 - 406

Zemanick ET., Wagner BD., Robertson CE., Ahrens RC., Chmiel JF., Clancy JP., Gibson RL., Harris WT., Kurland G., Laguna TA., McColley SA., McCoy K., Retsch-Bogart G., Sobush KT., Zeitlin PL., Stevens MJ., Accurso FJ., Sagel SD., et al
Airway microbiota across age and disease spectrum in cystic fibrosis
European Respiratory Journal 2017; 50: 5:1700832

Zhou E., Seminara AB., Kim SK., Hall CL., Wang Y., Lee VT.
Thiol-benzo-triazolo-quinazolinone Inhibits Alg44 Binding to c-di-GMP and Reduces Alginate Production by Pseudomonas aeruginosa
ACS Chemical Biology 2017; 12: 3076 – 3085

Nutrition

Barni GC., Forte GC., Forgiarini LF., Abrahao CLD., Dalcin PDR.
Factors associated with malnutrition in adolescent and adult patients with cystic fibrosis
Jornal Brasileiro de Pneumologia 2017; 43: 337 - 343

Calvo-Lerma J., Martinez-Barona S., Masip E., Fornes V., Ribes-Koninckx C.
Pancreatic enzyme replacement therapy in cystic fibrosis: dose, variability and coefficient of fat absorption
Revista Espanola de Enfermedades Digestivas 2017; 109: 684 - 689

Coriati A., Labreche E., Mailhot M., Mircescu H., Berthiaume Y., Lavoie A., Lhoret RR.
Vitamin D-3 supplementation among adult patients with cystic fibrosis
Clinical Nutrition 2017; 36: 1580 - 1585

Hermes WA., Alvarez JA., Lee MJ., Chedsachai S., Lodin D., Horst R., Tangpricha V.
Prospective, Randomized, Double-Blind, Parallel-Group, Comparative Effectiveness Clinical Trial Comparing a Powder Vehicle Compound of Vitamin D with an Oil Vehicle Compound in Adults With Cystic Fibrosis
Journal of Parenteral and Enteral Nutrition 2017; 41: 952 - 958

Hollander FM., de Roos NM., Heijerman HGM.
The optimal approach to nutrition and cystic fibrosis: latest evidence and recommendations
Current Opinion in Pulmonary Medicine 2017; 23: 556 - 561

Hollander FM., de Roos NM., van Meerkirk GB., van Berkhouft FT., Heijerman HGM., van de Graaf EA.
Body Weight and Body Mass Index in Patients with End-Stage Cystic Fibrosis Stabilize After the Start of Enteral Tube Feeding
Journal of the Academy of Nutrition and Dietetics 2017; 117: 1808 - 1815

Kanhere M., Chassaing B., Gewirtz AT., Tangpricha V.
Role of vitamin D on gut microbiota in cystic fibrosis
Journal of Steroid Biochemistry and Molecular Biology 2018; 175: 82 - 87

Konstantinopoulou S., Tapia IE.
Vitamin D and the lung
Paediatric Respiratory Reviews 2017; 24: 39 - 43

Lambe C., Sermet I.
Nutritional management in cystic fibrosis
Correspondances en Métabolismes Hormones Diabète et Nutrition 2017; 21: 232 - 236

Mathyssen C., Gayan-Ramirez G., Bouillon R., Janssens W.
Vitamin D supplementation in respiratory diseases: evidence from randomized controlled trials
Polish Archives of Internal Medicine-Polskie Archiwum Medycyny 2017; 127: 775 - 784

Sapieka E., Krzyzanowska P., Walkowiak D., Wenska-Chyzy E., Szczepanik M., Cofta S., Pogorzelski A., Skorupa W., Walkowiak J.
Vitamin A status and its determinants in patients with cystic fibrosis
Acta Scientiarum Polonorum-Technologia Alimentaria 2017; 13: 345 – 354

Physiotherapy

Hassanzad M., Masouleh SK., Nejad ST., Karimzadeh S., Velayati AA.
Efficacy of Noninvasive Positive Pressure Ventilation for Improving the Respiratory Function, Use of Accessory Respiratory Muscles, Quality of Sleep and Nutrition of Cystic Fibrosis Patients
Iranian Journal of Pediatrics 2017; 27: 4:e14597

Manor E., Gur M., Geffen Y., Bentur L.
Cleaning and infection control of airway clearance devices used by CF patients
Chronic Respiratory Disease 2017; 14: 370 - 376

McCormack P., Burnham P., Southern KW.
Autogenic drainage for airway clearance in cystic fibrosis
Cochrane Database of Systematic Reviews 2017; : 10:CD009595

Ramsey KA., Foong RE., Gradosic J., Harper A., Skoric B., Clem C., Davis M., Turkovic L., Stick SM., Davis SD., Ranganathan SC., Hall GL.
Multiple-Breath Washout Outcomes Are Sensitive to Inflammation and Infection in Children with Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 1436 - 1442

Smith LJ., Macleod KA., Collier GI., Horn FC., Sheridan H., Aldag I., Taylor CJ., Cunningham S., Wild JM., Horsley A.
Supine posture changes lung volumes and increases ventilation heterogeneity in cystic fibrosis
PLoS One 2017; 12: 11:e0188275

Voldby C., Green K., Rosthoj S., Kongstad T., Philipsen L., Buchvald F., Skov M., Pressler T., Gustafsson P., Nielsen KG.
The effect of time-of-day and chest physiotherapy on multiple breath washout measures in children with clinically stable cystic fibrosis
PLoS One 2018; 13: 1:e0190894

Psychosocial

Eakin MN., Chung SE., Hoehn J., Borrelli B., Rand-Giovannetti D., Riekert KA.
Development and validation of CF-Medication Beliefs Questionnaire: A mixed-methods approach
Journal of Cystic Fibrosis 2017; 16: 637 - 644

Gibson F.
Review: Using poetry to explore normalcy as a coping mechanism for young people with cystic fibrosis
Journal of Research in Nursing 2017; 22: 492 - 493

Gorrie A., Archibald AD., Ioannou L., Curnow L., McClaren B.
Exploring approaches to facilitate family communication of genetic risk information after cystic fibrosis population carrier screening
Journal of Community Genetics 2018; 9: 71 - 80

Leroy S., Pradelli J.
Pathophysiology and current affairs in the management of cystic fibrosis
Correspondances en Métabolismes Hormones Diabète et Nutrition 2017; 21: 226 - 230

MacDonald K.
Using poetry to explore normalcy as a coping mechanism for young people with cystic fibrosis
Journal of Research in Nursing 2017; 22: 479 - 491

Moola FJ., Garcia E., Huynh E., Henry L., Penfound S., Consunji-Araneta R., Faulkner GEJ.
Physical Activity Counseling for Children With Cystic Fibrosis
Respiratory Care 2017; 62: 1466 - 1473

Vandekerckhove K., Keyzer M., Cornette J., Coomans I., Pyl F., De Baets F., Schelstraete P., Haerynck F., De Wolf D., Van Daele S., Boone J.
Exercise performance and quality of life in children with cystic fibrosis and mildly impaired lung function: relation with antibiotic treatments and hospitalization
European Journal of Pediatrics 2017; 176: 1689 - 1696

Wallenwein A., Schwarz M., Goldbeck L.
Quality of life among German parents of children with cystic fibrosis: the effects of being a single caregiver
Quality of Life Research 2017; 26: 3289 - 3296

Xie DX., Wu J., Kelly K., Brown RF., Shannon C., Virgin FW.
Evaluating the sinus and Nasal Quality of Life Survey in the pediatric cystic fibrosis patient population
International Journal of Pediatric Otorhinolaryngology 2017; 102: 133 - 137

Pulmonology

Aronsohn J., Dowling O., Kars M., Roseman A.
Massive hemoptysis during general endotracheal anesthesia in adults with Cystic Fibrosis
Journal of Clinical Anesthesia 2017; 42: 17 - 18

Burgel PR., Reid DW., Aaron SD.
A first step to STOP cystic fibrosis exacerbation
Journal of Cystic Fibrosis 2017; 16: 529 - 531

Chelabi R., Soumagne T., Guillien A., Puyraveau M., Degano B.
In cystic fibrosis, lung clearance index is sensitive to detecting abnormalities appearing at exercise in children with normal spirometry
Respiratory Physiology & Neurobiology 2018; 247: 9 - 11

Conrad DJ., Bailey BA., Hardie JA., Bakke PS., Eagan TML., Aarli BB.
Median regression spline modeling of longitudinal FEV1 measurements in cystic fibrosis (CF) and chronic obstructive pulmonary disease (COPD) patients
PLoS One 2017; 12: 12:e0190061

Davies G., Stocks J., Thia LP., Hoo AF., Bush A., Aurora P., Brennan L., Lee S., Lum S., Cottam P., Miles J., Chudleigh J., Kirkby J., Balfour-Lynn IM., Carr SB., Wallis C., Wyatt H., Wade A.

Pulmonary function deficits in newborn screened infants with cystic fibrosis managed with standard UK care are mild and transient

European Respiratory Journal 2017; 50: 5:1700326

DeBoer EM., Kroehl ME., Wagner BD., Accurso FJ., Harris JK., Lynch DA., Sagel SD., Deterding RR.
Proteomic profiling identifies novel circulating markers associated with bronchiectasis in cystic fibrosis
Proteomics Clinical Applications 2017; 11: 9-10:1600147

Dickerhof N., Pearson JF., Hoskin TS., Berry LJ., Turner R., Sly PD., Kettle AJ.

Oxidative stress in early cystic fibrosis lung disease is exacerbated by airway glutathione deficiency
Free Radical Biology and Medicine 2017; 113: 236 - 243

Espel JC., Palac HL., Cullina JF., Clarke AP., McColley SA., Prickett MH., Jain M.

Antibiotic duration and changes in FEV1 are not associated with time until next exacerbation in adult cystic fibrosis: a single center study

BMC Pulmonary Medicine 2017; 17: ArNo: 160

mcGunasekara L., Al-Saidy M., Green F., Pratt R., Bjornson C., Yang AL., Schoel WM., Mitchell I., Brindle M., Montgomery M., Keys E., Dennis J., Shrestha G., Amrein M.
Pulmonary surfactant dysfunction in pediatric cystic fibrosis: Mechanisms and reversal with a lipid-sequestering drug
Journal of Cystic Fibrosis 2017; 16: 565 - 572

Hoppe JE., Wagner BD., Sagel SD., Accurso FJ., Zemanick ET.
Pulmonary exacerbations and clinical outcomes in a longitudinal cohort of infants and preschool children with cystic fibrosis
BMC Pulmonary Medicine 2017; 17: ArNo: 188

Kieninger E., Yammie S., Korten I., Anagnostopoulou P., Singer F., Frey U., Mornand A., Zanolari M., Rochat I., Trachsel D., Mueller-Suter D., Moeller A., Casaulta C., Latzin P.

Elevated lung clearance index in infants with cystic fibrosis shortly after birth

European Respiratory Journal 2017; 50: 5:1700580

Konstantinidis I., Fotoulaki M., Iakovou I., Chatziavramidis A., Mpalaris V., Shobat K., Markou K.

Vitamin D-3 deficiency and its association with nasal polyposis in patients with cystic fibrosis and patients with chronic rhinosinusitis

American Journal of Rhinology & Allergy 2017; 31: 395 - 400

Krantz C., Janson C., Hollsing A., Alving K., Malinovschi A.
Exhaled and nasal nitric oxide in relation to lung function, blood cell counts and disease characteristics in cystic fibrosis
Journal of Breath Research 2017; 11: 2:026001

Kuo WY., de Bruijne M., Petersen J., Nasserinejad K., Ozturk H., Chen Y., Perez-Rovira A., Tiddens HAWM. Diagnosis of bronchiectasis and airway wall thickening in children with cystic fibrosis: Objective airway-artery quantification
European Radiology 2017; 27: 4680 - 4689

Kuo WY., Soffers T., Andrinopoulou ER., Rosenow T., Ranganathan S., Turkovic L., Stick SM., Tiddens HAWM.
Quantitative assessment of airway dimensions in young children with cystic fibrosis lung disease using chest computed tomography
Pediatric Pulmonology 2017; 52: 1414 - 1423

Lapierre SG., Phelippeau M., Hakimi C., Didier Q., Reynaud-Gaubert M., Dubus JC., Drancourt M.
Cystic fibrosis respiratory tract salt concentration An Exploratory Cohort Study
Medicine 2017; 96: 47:e8423

- Lechtnin N., Mayer-Hamblett N., West NE., Allgood S., Wilhelm E., Khan U., Aitken ML., Ramsey BW., Boyle MP., Mogayzel PJ., Gibson RL., Orenstein D., Milla C., Clancy JP., Antony V., Goss CH.**
 Home Monitoring of Patients with Cystic Fibrosis to Identify and Treat Acute Pulmonary Exacerbations eICE Study Results
American Journal of Respiratory and Critical Care Medicine 2017; 196: 1144 - 1151
- Li D., Keogh R., Clancy JP., Szczesniak RD.**
 Flexible semiparametric joint modeling: an application to estimate individual lung function decline and risk of pulmonary exacerbations in cystic fibrosis
Emerging Themes in Epidemiology 2017; 14: ArNo: 13
- McGarry ME., Neuhaus JM., Nielson DW., Burchard E., Ly NP.**
 Pulmonary function disparities exist and persist in Hispanic patients with cystic fibrosis: A longitudinal analysis
Pediatric Pulmonology 2017; 52: 1550 - 1557
- Olszowiec-Chlebna M., Trzcinski K., Stelmach I.**
 Massive nasal polyposis in a patient with newly diagnosed cystic fibrosis
Advances in Respiratory Medicine 2017; 85: 121 - 123
- Pittman JE., Wylie KM., Akers K., Storch GA., Hatch J., Quante J., Frayman KB., Clarke N., Davis M., Stick SM., Hall GL., Montgomery G., Ranganathan S., Davis SD., Ferkol TW.**
 Association of Antibiotics, Airway Microbiome, and Inflammation in Infants with Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 1548 - 1555
- Radtke T., Benden C., Maggi-Beba M., Kriemler S., van der Lee I., Dressel H.**
 Intra-session and inter-session variability of nitric oxide pulmonary diffusing capacity in adults with cystic fibrosis
Respiratory Physiology & Neurobiology 2017; 246: 33 - 38
- Ramsey KA., Foong RE., Grdasic J., Harper A., Skoric B., Clem C., Davis M., Turkovic L., Stick SM., Davis SD., Ranganathan SC., Hall GL.**
 Multiple-Breath Washout Outcomes Are Sensitive to Inflammation and Infection in Children with Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 1436 - 1442
- Rayment JH., Ratjen F.**
 Another Brick in the Wall: Lung Clearance Index and Lower Airways Pathology in Preschool Cystic Fibrosis
Annals of the American Thoracic Society 2017; 14: 1389 - 1390
- Sanders DB., Zhao QQ., Li ZH., Farrell PM.**
 Poor recovery from cystic fibrosis pulmonary exacerbations is associated with poor long-term outcomes
Pediatric Pulmonology 2017; 52: 1268 - 1275
- Schechter MS.**
 Comparing effectiveness and outcomes in asthma and cystic fibrosis
Paediatric Respiratory Reviews 2017; 24: 24 - 28
- Schultz A., Puvvadi R., Borisov SM., Shaw NC., Klimant I., Berry LI., Montgomery ST., Nguyen T., Kreda SM., Kicic A., Noble PB., Button B., Stick SM.**
 Airway surface liquid pH is not acidic in children with cystic fibrosis
Nature Communications 2017; 8: ArNo: 1409
- Sellers ZM., Illek B., Figueira MF., Hari G., Joo NS., Sibley E., Souza-Menezes J., Morales MM., Fischer H., Wine JJ.**
 Impaired PGE(2)-stimulated Cl- and HCO3- secretion contributes to cystic fibrosis airway disease
PLoS One 2017; 12: e0189894
- van Horck M., Winkens B., Wesseling G., van Vliet D., van de Kant K., Vaassen S., de Winter-de Groot K., de Vreede I., Jobsis Q., Dompeling E.**
 Early detection of pulmonary exacerbations in children with Cystic Fibrosis by electronic home monitoring of symptoms and lung function
Scientific Reports 2017; 7: ArNo: 12350
- VanDevanter DR., Heltshe SL., Spahr J., Beckett VV., Daines CL., Dasenbrook EC., Gibson RL., Jain R., Sanders DB., Bc CHG., Flume PA.**
 Rationalizing endpoints for prospective studies of pulmonary exacerbation treatment response in cystic fibrosis
Journal of Cystic Fibrosis 2017; 16: 607 - 615
- Virgin FW.**
 Clinical Chronic Rhinosinusitis Outcomes in Pediatric Patients with Cystic Fibrosis
Laryngoscope Investigative Otolaryngology 2017; 2: 276 - 280
- Wandro S., Carmody L., Gallagher T., LiPuma JJ., Whiteson K.**
 Making It Last: Storage Time and Temperature Have Differential Impacts on Metabolite Profiles of Airway Samples from Cystic Fibrosis Patients
Msystems 2017; 2: 6:UNSP e00100-
- Weber SAT., Iyomasa RM., Correa CD., Florentino WNM., Ferrari GF.**
 Nasal polyposis in cystic fibrosis: follow-up of children and adolescents for a 3-year period
Brazilian Journal of Otorhinolaryngology 2017; 83: 677 - 682
- West NE., Beckett VV., Jain R., Sanders DB., Nick JA., Heltshe SL., Dasenbrook EC., VanDevanter DR., Solomon GM., Goss CH., Flume PA.**
 Standardized Treatment of Pulmonary Exacerbations (STOP) study: Physician treatment practices and outcomes for individuals with cystic fibrosis with pulmonary Exacerbations
Journal of Cystic Fibrosis 2017; 16: 600 - 606
- Williams CA., Tomlinson OW., Chubbsack LV., Stevens D., Saynor ZL., Oades PJ., Barker AR.**
 The oxygen uptake efficiency slope is not a valid surrogate of aerobic fitness in cystic fibrosis
Pediatric Pulmonology 2018; 53: 36 - 42
- Woodley FW., Moore-Clingenpeel M., Machado RS., Nemastil CJ., Jadcherla SR., Hayes D., Kopp BT., Kaul A., Di Lorenzo C., Mousa H.**
 Not All Children with Cystic Fibrosis Have Abnormal Esophageal Neutralization during Chemical Clearance of Acid Reflux
Pediatric Gastroenterology Hepatology & Nutrition 2017; 20: 153 - 159

Radiology

- Braun C., Bacchetta J., Braillon P., Chapurlat R., Drai J., Reix P.**
 Children and adolescents with cystic fibrosis display moderate bone microarchitecture abnormalities: data from high-resolution peripheral quantitative computed tomography
Osteoporosis International 2017; 28: 3179 - 3188
- Gauthier R., Cabon Y., Giroux-Metges MA., Du Boisbaudry C., Reix P., Le Bourgeois M., Chiron R., Molinari N., Saguintaa M., Amsalem F., Matecki S.**
 Early follow-up of lung disease in infants with cystic fibrosis using the raised volume rapid thoracic compression technique and computed tomography during quiet breathing
Pediatric Pulmonology 2017; 52: 1283 - 1290

Kaireit TF., Sorrentino SA., Renne J., Schoenfeld C., Voskrebenev A., Gutberlet M., Schulz A., Jakob PM., Hansen G., Wacker F., Welte T., Tummller B., Vogel-Claussen J.
Functional lung MRI for regional monitoring of patients with cystic fibrosis
PLoS One 2017; 12: e0187483

Kuo WY., Soffers T., Andrinopoulou ER., Rosenow T., Ranganathan S., Turkovic L., Stick SM., Tiddens HAWM.
Quantitative assessment of airway dimensions in young children with cystic fibrosis lung disease using chest computed tomography
Pediatric Pulmonology 2017; 52: 1414 - 1423

Nyilas S., Bauman G., Sommer G., Stranzinger E., Pusterla O., Frey U., Korten I., Singer F., Casaulta C., Bieri O., Latzin P.
Novel magnetic resonance technique for functional imaging of cystic fibrosis lung disease
European Respiratory Journal 2017; 50: 6:1701464

Schaefer JF., Hector A., Schmidt K., Teufel M., Fleischer S., Graepler-Mainka U., Riethmueller J., Gatidis S., Schaefer S., Nikolaou K., Hartl D., Tsiflikas I.

A semiquantitative MRI-Score can predict loss of lung function in patients with cystic fibrosis: Preliminary results
European Radiology 2018; 28: 74 - 84

Tabori H., Jaudszus A., Arnold C., Mentzel HJ., Lorenz M., Michl RK., Lehmann T., Renz DM., Mainz JG.
Relation of Ultrasound Findings and Abdominal Symptoms obtained with the CFAbd-Score in Cystic Fibrosis Patients
Scientific Reports 2017; 7: ArNo: 17465

Ward R., Carroll WD., Cunningham P., Ho SA., Jones M., Lenney W., Thompson D., Gilchrist FJ.
Radiation dose from common radiological investigations and cumulative exposure in children with cystic fibrosis: an observational study from a single UK centre
BMJ Open 2017; 7: 8:e017548

Screening

Currier RJ., Sciortino S., Liu RL., Bishop T., Koupaei RA., Feuchtabaum L.
Genomic sequencing in cystic fibrosis newborn screening: what works best, two-tier predefined CFTR mutation panels or second-tier CFTR panel followed by third-tier sequencing?
Genetics in Medicine 2017; 19: 1159 - 1163

Esteves CZ., Dias LD., Lima ED., de Oliveira DN., Melo CFOR., Delafiori J., Gomez CCS., Ribeiro JD., Ribeiro AF., Levy CE., Catharino RR.
Skin Biomarkers for Cystic Fibrosis: A Potential Non-Invasive Approach for Patient Screening
Frontiers in Pediatrics 2018; 5: ArNo: 290

Gorrie A., Archibald AD., Ioannou L., Curnow L., McLaren B.
Exploring approaches to facilitate family communication of genetic risk information after cystic fibrosis population carrier screening
Journal of Community Genetics 2018; 9: 71 - 80

Hayeems RZ., Miller FA., Vermeulen M., Potter BK., Chakraborty P., Davies C., Carroll JC., Ratjen F., Guttmann A.
False-Positive Newborn Screening for Cystic Fibrosis and Health Care Use
Pediatrics 2017; 140: 5:e20170604

Icke S., Genc RE.
National Newborn Screening Tests Carried Out with Heel Lance and Their Importance
Journal of Pediatric Research 2017; 4: 186 - 190

Solomon GM., Liu B., Sermet-Gaudelus I., Fajac I., Wilcanski M., Vermeulen F., Rowe SM.
A multiple reader scoring system for Nasal Potential Difference parameters
Journal of Cystic Fibrosis 2017; 16: 573 - 578

Ulph F., Wright S., Dharni N., Payne K., Bennett R., Roberts S., Walshe K., Lavender T.
Provision of information about newborn screening antenatally: a sequential exploratory mixed-methods project
Health Technology Assessment 2017; 21: 1

Therapy

Ayoub F., Lascano J., Morelli G.
Proton Pump Inhibitor Use Is Associated with an Increased Frequency of Hospitalization in Patients With Cystic Fibrosis
Gastroenterology Research 2017; 10: 288 - 293

Bulloch MN., Hanna C., Giovane R.
Lumacaftor/ivacaftor, a novel agent for the treatment of cystic fibrosis patients who are homozygous for the F508del CFTR mutation
Expert Review of Clinical Pharmacology 2017; 10: 1055 - 1072

Dagan A., Cohen-Cymberknob M., Shtenberg M., Levine H., Vilozni D., Bezalel Y., Bat-El Bar Aluma., Sarouk I., Ashkenazi M., Lavie M., Tsabari R., Blau H., Kerem E., Bentur L., Efrati O., Livnat G.
Ivacaftor for the p.Ser549Arg (S549R) gating mutation - The Israeli experience
Respiratory Medicine 2017; 131: 225 - 228

De Boeck K., Davies JC.
Where are we with transformational therapies for patients with cystic fibrosis?
Current Opinion in Pharmacology 2017; 34: 70 - 75

Dilokthornsakul P., Patidar M., Campbell JD.
Forecasting the Long-Term Clinical and Economic Outcomes of Lumacaftor/Ivacaftor in Cystic Fibrosis Patients with Homozygous phe508del Mutation
Value in Health 2017; 20: 1329 - 1335

Dryden C., Wilkinson J., Young D., Brooker RJ.
The impact of 12 months treatment with ivacaftor on Scottish paediatric patients with cystic fibrosis with the G551D mutation: a review
Archives of Disease in Childhood 2018; 103: 68 - 70

Guevara MT., McColley SA.
The safety of lumacaftor and ivacaftor for the treatment of cystic fibrosis
Expert Opinion On Drug Safety 2017; 16: 1305 - 1311

Guimbellot J., Sharma J., Rowe SM.
Toward inclusive therapy with CFTR modulators: Progress and challenges
Pediatric Pulmonology 2017; 52:

Hanrahan JW., Matthes E., Carlile G., Thomas DY.
Corrector combination therapies for F508del-CFTR
Current Opinion in Pharmacology 2017; 34: 105 - 111

Jennings MT., Dezube R., Paranjape S., West NE., Hong G., Braun A., Grant J., Merlo CA., Lechtzin N.
An Observational Study of Outcomes and Tolerances in Patients with Cystic Fibrosis Initiated on Lumacaftor/Ivacaftor
Annals of the American Thoracic Society 2017; 14: 1662 - 1666

Lingam S., Thonghin N., Ford RC.
Investigation of the effects of the CFTR potentiator ivacaftor on human P-glycoprotein (ABCB1)
Scientific Reports 2017; 7: ArNo: 17481

- Maiuri L., Raia V., Kroemer G.**
Strategies for the etiological therapy of cystic fibrosis
Cell Death and Differentiation 2017; 24: 1825 - 1844
- Oliver KE., Han ST., Sorscher EJ., Cutting GR.**
Transformative therapies for rare CFTR missense alleles
Current Opinion in Pharmacology 2017; 34: 76 - 82
- Payne JE., Dubois AV., Ingram RJ., Weldon S., Taggart CC., Elborn JS., Tunney MM.**
Activity of innate antimicrobial peptides and ivacaftor against clinical cystic fibrosis respiratory pathogens
International Journal of Antimicrobial Agents 2017; 50: 427 - 435
- Rowe SM., Daines C., Ringshausen FC., Kerem E., Wilson J., Tullis E., Nair N., Simard C., Han L., Ingenito EP., McKee C., Lekstrom-Himes J., Davies JC.**
Tezacaftor-Ivacaftor in Residual-Function Heterozygotes with Cystic Fibrosis
New England Journal of Medicine 2017; 377: 2024 - 2035
- Rowe SM., McColley SA., Rietschel E., Li XL., Bell SC., Konstan MW., Marigowda G., Waltz D., Boyle MP.**
Lumacaftor/Ivacaftor Treatment of Patients with Cystic Fibrosis Heterozygous for F508del-CFTR
Annals of the American Thoracic Society 2017; 14: 213 - 219
- Schneider EK., Reyes-Ortega F., Li J., Velkov T.**
Optimized HPLC-MS Method for the High-Throughput Analysis of Clinical Samples of Ivacaftor, Its Major Metabolites, and Lumacaftor in Biological Fluids of Cystic Fibrosis Patients
JOVE-journal of Visualized Experiments 2017; : 128:e56084
- Scott DW., Walker MP., Sesma J., Wu B., Stuhlmiller TJ., Sabater JR., Abraham WM., Crowder TM., Christensen DJ., Tarhan R.**
SPX-101 Is a Novel Epithelial Sodium Channel-targeted Therapeutic for Cystic Fibrosis That Restores Mucus Transport
American Journal of Respiratory and Critical Care Medicine 2017; 196: 734 - 744
- Smiths S., Edwards CT.**
Long-acting inhaled bronchodilators for cystic fibrosis
Cochrane Database of Systematic Reviews 2017; : 12:CD012102
- Solomon GM., Fu LW., Rowe SM., Collawn JF.**
The therapeutic potential of CFTR modulators for COPD and other airway diseases
Current Opinion in Pharmacology 2017; 34: 132 - 139
- Taylor-Cousar JL., Munck A., McKone EF., van der Ent CK., Moeller A., Simard C., Wang LT., Ingenito EP., McKee C., Lu YM., Lekstrom-Himes J., Elborn JS.**
Tezacaftor-Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508del
New England Journal of Medicine 2017; 377: 2013 - 2023
- VanDevanter DR., Craib ML., Pasta DJ., Millar SJ., Morgan WJ., Konstan MW.**
Cystic fibrosis clinical characteristics associated with dornase alfa treatment regimen change
Pediatric Pulmonology 2018; 53: 43 - 49
- VanDevanter DR., Mayer-Hamblett N.**
Innovating cystic fibrosis clinical trial designs in an era of successful standard of care therapies
Current Opinion in Pulmonary Medicine 2017; 23: 530 - 535
- Walker MP., Cowlen M., Christensen D., Miyamoto M., Barley P., Crowder T.**
Nonclinical safety assessment of SPX-101, a novel peptide promoter of epithelial sodium channel internalization for the treatment of cystic fibrosis
Inhalation Toxicology 2017; 29: 356 - 365
- Zeitlin PL., Diener-West M., Callahan KA., Lee S., Talbot CC., Pollard B., Boyle MP., Lechtzin N.**
Digitoxin for Airway Inflammation in Cystic Fibrosis: Preliminary Assessment of Safety, Pharmacokinetics, and Dose Finding
Annals of the American Thoracic Society 2017; 14: 220 - 229
- ## Transplantation
- Crawford TC., Magruder JT., Grimm JC., Suarez-Pierre A., Zhou X., Ha JS., Higgins RS., Broderick SR., Orens JB., Shah P., Merlo CA., Kim BS., Bush EL.**
Impaired Renal Function Should Not Be a Barrier to Transplantation in Patients with Cystic Fibrosis
Annals of Thoracic Surgery 2017; 104: 1231 - 1236
- Dellon E., Goldfarb SB., Hayes D., Sawicki GS., Wolfe J., Boyer D.**
Pediatric lung transplantation and end of life care in cystic fibrosis: Barriers and successful strategies
Pediatric Pulmonology 2017; 52:
- Dupont L.**
Lung transplantation in cystic fibrosis patients with difficult to treat lung infections
Current Opinion in Pulmonary Medicine 2017; 23: 574 - 579
- Kennedy CC., Razonable RR.**
Fungal Infections After Lung Transplantation
Clinics in Chest Medicine 2017; 38: 511 - +
- Lazor T., Grasemann H., Solomon M., Anthony SJ.**
Quality of life outcomes following pediatric lung transplantation
Pediatric Pulmonology 2017; 52: 1495 - 1501
- Levine H., Prais D., Raviv Y., Rusanov V., Rosengarten D., Saute M., Hoshen M., Mussaffi H., Blau H., Kramer MR.**
Lung transplantation in cystic fibrosis patients in Israel: The importance of ethnicity and nutritional status
Clinical Transplantation 2017; 31: 11:e13111
- Markelic I., Jakopovic M., Klepetko W., Dzubur F., Hecimovic A., Makek MJ., Samarzija M., Dugac AV.**
Lung Abscess: An Early Complication of Lung Transplantation in a Patient with Cystic Fibrosis
International Journal of Organ Transplantation Medicine 2017; 8: 213 - 216
- Noh SR., Lee E., Yoon J., Jung S., Yang SI., Yu J., Hong SJ.**
The First Successful Lung Transplantation in a Korean Child with Cystic Fibrosis
Journal of Korean Medical Science 2017; 32: 2073 - 2078
- Parize P., Boussaud V., Poinsignon V., Sitterle E., Botterel F., Lefevre S., Guillemain R., Dannaoui E., Billaud EM.**
Clinical outcome of cystic fibrosis patients colonized by *Scedosporium* species following lung transplantation: A single-center 15-year experience
Transplant Infectious Disease 2017; 19: 5:e12738
- Snell G., Reed A., Stem M., Hadjiliadis D.**
The evolution of lung transplantation for cystic fibrosis: A 2017 update
Journal of Cystic Fibrosis 2017; 16: 553 - 564
- ## Urology
- Frayman KB., Kazmerski TM., Sawyer SM.**
A systematic review of the prevalence and impact of urinary incontinence in cystic fibrosis
Respirology 2018; 23: 46 - 54
- Santoro D., Postorino A., Lucanto C., Costa S., Cristadoro S., Pellegrino S., Conti G., Buemi M., Magazzu G., Bellinghieri G.**
Cystic Fibrosis: A Risk Condition for Renal Disease
Journal of Renal Nutrition 2017; 27: 470 - 473