Dear Friends,

I hope you had a good start of the year.

2014 was another very busy year for the European Cystic Fibrosis Society, with not only the Basic Science Conference in Malta and the Annual conference in Gothenburg, but a number of special projects and Working Group meetings were held and some selected updates are given below.

Later in this newsletter, you will find some new initiatives recently introduced which comprises the creation of an ECFS Cystic Fibrosis Molecular & Cell Biology and Physiology Basic Science Working Group, ECFS Short-Term Fellowships and a new possibility for long term membership.

There will be Board elections in 2015 with both Dominique Hubert and Milan Macek ending their terms in office. 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. The Board is now predominantly Pediatric Physicians so I encourage Adult Physicians and Nurses and AHP to please consider standing. 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 2015 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. We have excellent programmes 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,

Stuart Elborn, ECFS President
2. 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 10 June 2015 of the annual conference in Brussels. You are cordially invited to nominate a candidate for this award. The deadline for proposals is 15 March 2015. 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).

3. Gerd Döring Award - Call for Nominations

The Gerd Döring Award is a new initiative of the European Cystic Fibrosis Society and will be 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 Brussels. The award will be judged primarily (80%) on a paper published in the previous 3 calendar years (2012-2014) 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 15 March 2015. 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).

4. ECFS Board Elections - Call for Nominations

The ECFS cordially invites nominations for the following Board positions. In June 2015, Stuart Elborn will finish his term as President and Kris De Boeck, currently President elect, will fully take over the role. Dominique Hubert will finish her term on the Board in June, and Milan Macek is stepping down from the Board after completing two terms. Job Descriptions and person specifics are available here.

Nominations should be sent to the ECFS Executive Director Christine Dubois (christine.dubois@ecfs.eu) by 18 March 2015 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.

5. CTN Steering Committee meeting Barcelona, January 29-30

Investigators from 28 of the 30 CTN sites were able to join this yearly winter meeting. Patient organizations from Belgium, Germany, Italy, Luxemburg, Switzerland, The Netherlands and UK were also represented. As in previous years, Dr. George Retsch-Bogart traveled to Europe and attended as CFF-TDN representative.
Dr. Tim Lee (Leeds) is now the ECFS-CTN Director for the coming 3 years, replacing Prof. Isabelle Fajac. Isabelle was thanked for her excellent leadership and for having moved the network forward during her term.

3 more people ended their term in the Executive Committee and were thanked for all their valuable work and input: Pavel Drejinek (Prague), Nico Derichs (Germany) and Andreas Reimann (patient organization representative, Germany). This also means that new members have joined the Executive Committee: Damian Downey (Belfast), Lieven Dupont (Leuven) and Michael Fayon (Bordeaux) as investigators and Paola De Carli from the French patient organisation.

The first day of the steering committee meeting looked back at the 2014 activities, but mainly focused on the business plan for the upcoming 3 years. The ECFS-CTN wants to increase its impact on CF clinical trials in Europe. Some important topics are the involvement of patients and the establishment of an advisory group of young persons with CF. Also the ECFS-CTN wants to develop an Investigator Initiated Trial involving all stakeholders from the start. Of course, the work on new outcome parameters and surrogate endpoints will be continued.

During the 2 days, updates were provided by the different committees (protocol review, training, DSMB, standardization). The yearly report on site quality assessment was presented and strategies to cope with the pipeline of upcoming trials were discussed.

Additional meetings were organized between the Executive Committees of the ECFS-CTN and the ECFS Patient Registry and with the patient organizations funding research. As always the ECFS winter meeting has been an excellent opportunity for all parties to meet each other, to have fruitful discussions and to work on new initiatives.

6. Patient Registry Executive and Steering Committees meetings

The country representatives and the staff of the ECFS Patient Registry met to discuss current developments and future plans. The registry keeps expanding and centres of Lithuania, Macedonia, Romania and Ukraine were welcomed in 2014, which brings the number of participating countries to 28. In an animated atmosphere, topics such as the data-collection software ECFSTracker and its launch in Europe in the past year, the quality of data, the publication of the next annual report with 2013 data planned for January 2016, and the business plan were discussed, resulting in constructive input to take forward into the next months.

7. ECFS Cystic Fibrosis Molecular & Cell Biology and Physiology Basic Science Working Group

It is always a pleasure to announce the creation of a new working Group. This working group aims to establish solid grounds for Molecular & Cell Biology and Physiology of Cystic Fibrosis.

Margarida Amaral (PT) and Marcus Mall are respectively the Coordinator and Vice-coordinator of this new working group.

Any ECFS member can join the Working Group.

More information on the Working Group can be found here

The first meeting of the working group is planned on 25 March 2015, 09:30-12:00, in Albufeira, Portugal, before the opening of the 12th ECFS Basic Science Conference.
8. Clinical training fellowships

The ECFS has partnered with the course ‘Managing the care of children and adults with cystic fibrosis’ held twice a year in London, UK or on-line, www.cfcourse.co.uk, to offer clinical placements to members of the CF multidisciplinary team.

Sponsorship includes free attendance at the course in London or participation in the on-line course, followed by a two week placement at the Royal Brompton Hospital (paediatric or adult departments). Travel and accommodation will be covered.

The course is held twice a year in March and September, the on-line course can be done at any time: www.cfcourse.co.uk

If you are interested please contact the ECFS, info@ecfs.eu

9. Three year ECFS Special membership offer

This year a new membership offer was introduced: a 3 year membership for 300 € instead of 360 € (3*120€), giving access to the JCF, voting rights and reductions to the ECFS conferences. You can now show your faithfulness to the ECFS by subscribing for 3 years (2015, 16 and 17) in one step. So far, we are happy to report that this subscription type seems to be appreciated and quite a few members have chosen this new option.

10. Upcoming Events

- ECFS 12th Basic Science Conference - 25-28 March 2015 - Albufeira, Portugal
- ECFS Board Meeting - 09 June 2015 - Brussels, Belgium
- ECFS Quality Management Course, 09 June - Brussels, Belgium
- ECFS CF Course, 10 June - Brussels, Belgium
- 38th European CF Conference - 10-13 June 2015 - Brussels, Belgium

11. Current references in Cystic Fibrosis

Please scroll down to next page
CF References

Adults & Adolescents

Becher C., Regamey N., Spychiger E.
Transition - how adolescents with cystic fibrosis and their parents experience the change from paediatric to adult care
Pflege 2014; 27: 359 - 368

Harness-Brumley CL., Elliott AC., Rosenbluth DB., Raghavan D., Jain R.
Gender Differences in Outcomes of Patients with Cystic Fibrosis
Journal of Womens Health 2014; 23: 1012 - 1020

Horsley A., Siddiqui S.
Putting lung function and physiology into perspective: cystic fibrosis in adults
Respirology 2015; 20: 33 - 45

Hulzebos EHJ., Bomhof-Roordink H., van de Weert-van Leeuwen PBV., Twisk JWR., Artes HGM., van der Ent CK., Takken T.
Prediction of Mortality in Adolescents with Cystic Fibrosis
Medicine and Science In Sports and Exercise 2014; 46: 2047 - 2052

Oliver KN., Free ML., Bok C., McCoy KS., Lemanek KL., Emery CF.
Stigma and optimism in adolescents and young adults with cystic fibrosis
Journal of Cystic Fibrosis 2014; 13: 737 - 744

Patel EM., Swamy GK., Heine RP., Kuller JA., James AH., Grotegut CA.
Medical and obstetric complications among pregnant women with cystic fibrosis
American Journal of Obstetrics and Gynecology 2015; 212: 1:98.e1

Reverri EJ., Morrissey BM., Cross CE., Steinberg FM.
Inflammation, oxidative stress, and cardiovascular disease risk factors in adults with cystic fibrosis
Free Radical Biology and Medicine 2014; 76: 261 - 277

Reynolds N., Mrug S., Britton L., Guion K., Wolfe K., Gutierrez H.
Spiritual coping predicts 5-year health outcomes in adolescents with cystic fibrosis
Journal of Cystic Fibrosis 2014; 13: 593 - 600

VanWort TA., Lee JA., Karvir H., Whitehouse MC., Beim PY., Copperman AB.
Female cystic fibrosis mutation carriers and assisted reproductive technology: does carrier status affect reproductive outcomes?
Fertility and Sterility 2014; 102:

Yefet E., Salim R., Chazan B., Akel H., Romano S., Nachum Z.
The Safety of Quinolones in Pregnancy
Obstetrical & Gynecological Survey 2014; 69: 681 - 694

Animal Model

Cmielewski P., Donnelley M., Parsons DW.
Long-term therapeutic and reporter gene expression in lentiviral vector treated cystic fibrosis mice

Jaecklin T., Duerr J., Huang H., Rafii M., Bear CE., Ratjen F., Pencharz P., Kavanagh BP., Mall MA., Grasemann H.
Lung arginase expression and activity is increased in cystic fibrosis mouse models

Mokhtar HM., Giribabu N., Kassim N., Muniandy S., Salleh N.
Testosterone decreases fluid and chloride secretions in the uterus of adult female rats via down-regulating cystic fibrosis transmembrane regulator (CFTR) expression and functional activity
Journal Of Steroid Biochemistry And Molecular Biology 2014; 144: 361 - 372

Ng HP., Zhou Y., Song KJ., Hodges CA., Drumml ML., Wang GS.
Neutrophil-Mediated Phagocytic Host Defense Defect in Myeloid Cbf-Infected Mice

Antimicrobials

Bandara HMHN., Harb A., Kolacny D., Martins P., Smyth HDC.
Sound Waves Effectively Assist Tobramycin in Elimination of Pseudomonas aeruginosa Biofilms In vitro
AAPS PharmScitech 2014; 15: 1544 - 1564

Beloin C., Renard S., Ghigo JM., Lebeaux D.
Novel approaches to combat bacterial biofilms
Current Opinion In Pharmacology 2014; 18: 61 - 68

Berkhout MC., van Velzen AJ., Touw DJ., de Kok BM., Fokkens WJ., Heijerman HGM.
Systemic absorption of nasally administered tobramycin and colistin in patients with cystic fibrosis
Journal of Antimicrobial Chemotherapy 2014; 69: 3112 - 3115

Betegnie AL., Cracowski C., Bedouch P., Segond C., Robein-Dobrenez MJ., Pin I., Allenet B.
Peripherally inserted central catheter antibiotic therapy for cystic fibrosis patients
Revue des Maladies Respiratoires 2014; 31: 822 - 830

Dalhoff A.
Pharmacokinetics and Pharmacodynamics of Aerosolized Antibacterial Agents in Chronically Infected Cystic Fibrosis Patients
Clinical Microbiology Reviews 2014; 27: 753 - 782

Dosler S., Karaaslan E.
Inhibition and destruction of Pseudomonas aeruginosa biofilms by antibiotics and antimicrobial peptides
Peptides 2014; 62: 32 - 37

Du J., El-Sherbiny IM., Smyth HD.
Swellable Ciprofloxacin-Loaded Nano-in-Micro Hydrogel Particles for Local Lung Drug Delivery
AAPS PharmScitech 2014; 15: 1535 - 1544

Elkhatib W., Noreddin A.
Efficacy of Ciprofloxacin-Clinithromycin Combination Against Drug-Resistant Pseudomonas aeruginosa Mature Biofilm Using In Vitro Experimental Model
Microbial Drug Resistance 2014; 20: 575 - 582

GI M., Jeong J., Lee K., Lee KM., Toyofuku M., Yong DE., Yoon SS., Choi JY.
A Drug-Repositioning Screening Identifies Pentetic Acid as a Potential Therapeutic Agent for Suppressing the Elastase-Mediated Virulence of Pseudomonas aeruginosa
Antimicrobial Agents and Chemistry 2014; 58: 7205 - 7214

Habash MB., Park AJ., Vis EC., Harris RJ., Khursigara CM.
Synergy of Silver Nanoparticles and Aztreonam against Pseudomonas aeruginosa PA01 Biofilms
Antimicrobial Agents and Chemistry 2014; 58: 5818 - 5830

Harrison MJ., McCarthy M., Fleming C., Hickey C., Shortt C., Eustace JA., Murphy DM., Plant BJ.
Inhaled versus nebulised tobramycin: A real world comparison in adult cystic fibrosis (CF)
Lee JY., Chung ES., Na YJ., Kim H., Shin D., Ko KS.
Development of colistin resistance in pmrA-, phoP-, parK- and
cppR-inactivated mutants of Pseudomonas aeruginosa
Journal of Antimicrobial Chemotherapy 2014; 69: 2966 - 2971

Lehmann S., Pfannenstiel C., Friedrichs F., Kroger K.,
Wagner N., Tenbrock K.
Omalizumab: a new treatment option for allergic
bronchopulmonary aspergillosis in patients with cystic fibrosis
Therapeutic Advances In Respiratory Disease 2014; 8: 141 - 149

Lescar J., Meyer I., Akshita K., Srinivasaraghavan K.,
Verma C., Palous M., Mazier D., Datry A., Fekkar A.
Aspergillus fumigatus harbouring the sole Y121F mutation shows
decreased susceptibility to voriconazole but maintained
susceptibility to itraconazole and posaconazole
Journal of Antimicrobial Chemotherapy 2014; 69: 3244 - 3247

Lora-Tamayo J., Murillo O., Bergen PJ., Nation RL., Poudyal A.,
Luo XL., Yu HY., Ariza J., Li J.
Activity of colistin combined with doripenem at clinically relevant
concentrations against multidrug-resistant Pseudomonas aeruginosa
in an in vitro dynamic biofilm model
Journal Of Antimicrobial Chemotherapy 2014; 69: 2434 - 2442

Macia MD., Rojo-Moliner E., Oliver A.
Antimicrobial susceptibility testing in biofilm-growing bacteria
Clinical Microbiology And Infection 2014; 20: 981 - 990

Malott RJ., Wu CH., Lee TD., Hird TJ., Dalleska NF., Zlosnik JEA.,
Newman DK., Speert DP.
Fosmidomycin Decreases Membrane Hapenoids and Potentiates the
Effects of Colistin on Burkholderia multivorans Clinical Isolates
Antimicrobial Agents and Chemotherapy 2014; 58: 5211 - 5219

Mazurek H., Chiron R., Kucerova T., Geidel C., Bolbas K.,
Chuchalin A., Blanco-Aparicio M., Santoro D., Varoli G.,
Zibellini M., Cicirello HG., Antipkin YG.
Long-Term Efficacy and Safety of Aerosolized Tobramycin 300
mg/4 ml in Cystic Fibrosis
Pediatric Pulmonology 2014; 49: 1076 - 1089

Moreira AS., Silva D., Ferreira AR., Delgado L.
Antifungal treatment in allergic bronchopulmonary aspergillosis with
and without cystic fibrosis: a systematic review
Clinical and Experimental Allergy 2014; 44: 1210 - 1227

Okusanya OO., Bhavnani SM., Hammel JP., Forrest A.,
Aparicio M., Santoro D., Varoli G.,
Molenda N., Urbanova K., Weiser N., Kusche-Gullberg M.,
Janssens H.M., Pang PW., Lahap K., Pruliere E., Coste A.,
Edelman A.
Proteomic Analysis of Nasal Epithelial Cells from Cystic Fibrosis
Patients

Carbone A., Castellani S., Favia M., Diana A., Paracchini V.,
Di Gioia S., Seia M., Casavola V., Colombo C., Conese M.
Correction of defective CFTR/ENaC function and tightness of cystic
fibrosis airway epithelium by amniotic mesenchymal stromal (stem)
cells
Journal of Cellular and Molecular Medicine 2014; 18: 1631 - 1643

Jeanson L., Guerrera IC., Papon JF., Chhuon C., Zadigue P.,
Pruliere-Escabasse V., Amselem S., Escudier E., Coste A.,
Tinaut S., Lescar J., Meyer I., Akshita K., Fekkar A.
Antibiotic resistance in Pseudomonas aeruginosa biofilms: Towards
the development of novel anti-biofilm therapies
Journal of Biotechnology 2014; 191: 121 - 130

Tiddens HAWM., Bos AC., Malfroot A.
How Long Should We Maintain Long-Term Azithromycin Treatment
in Cystic Fibrosis Patients?
Pediatric Pulmonology 2015; 50: 103 – 104

Strempel N., Strehmel J., Overhage J.
Potential Application of Antimicrobial Peptides in the Treatment of
Bacterial Biofilm Infections

Taylor PK., Yeung ATY., Hancock REW.
Antibiotic resistance in Pseudomonas aeruginosa biofilms: Towards
the development of novel anti-biofilm therapeutics
Journal of Biotechnology 2014; 191: 121 - 130

Utley L., Tappenden P.
Dry powder inhalers in cystic fibrosis: same old drugs but different
benefits?
Current Opinion In Pulmonary Medicine 2014; 20: 607 - 612

Cell Biology

Carbone A., Castellani S., Favia M., Diana A., Paracchini V.,
Di Gioia S., Seia M., Casavola V., Colombo C., Conese M.
Correction of defective CFTR/ENaC function and tightness of cystic
fibrosis airway epithelium by amniotic mesenchymal stromal (stem)
cells
Journal of Cellular and Molecular Medicine 2014; 18: 1631 - 1643

Jeanson L., Guerrera IC., Papon JF., Chhuon C., Zadigue P.,
Pruliere-Escabasse V., Amselem S., Escudier E., Coste A.,
Tinaut S., Lescar J., Meyer I., Akshita K., Fekkar A.
Antibiotic resistance in Pseudomonas aeruginosa biofilms: Towards
the development of novel anti-biofilm therapies
Journal of Biotechnology 2014; 191: 121 - 130

Tiddens HAWM., Bos AC., Malfroot A.
How Long Should We Maintain Long-Term Azithromycin Treatment
in Cystic Fibrosis Patients?
Pediatric Pulmonology 2015; 50: 103 – 104
Voisin G., Bouvet GF., Legendre P., Dagenais A., Masse C., Berthiaume Y. Oxidative stress modulates the expression of genes involved in cell survival in Delta F508 cystic fibrosis airway epithelial cells Physiological Genomics 2014; 46: 634 - 646


CFTR

Amacher JF., Zhao RZ., Spaller MR., Madden DR. Chemically Modified Peptide Scaffolds Target the CFTR-Associated Ligand PDZ Domain PLoS One 2014; 9: e8103650


Chan HC., Jiang XH., Ruan YC. Emerging role of cystic fibrosis transmembrane conductor regulator as an epigenetic regulator: linking environmental cues to microRNAs Clinical and Experimental Pharmacology and Physiology 2014; 41: 615 - 622


El Hiani Y., Linsdell P. Metal Bridges Illuminate Transmembrane Domain Movements during Gating of the Cystic Fibrosis Transmembrane Conductance Regulator Chloride Channel Journal of Biological Chemistry 2014; 289: 28149 - 28159


Gosalia N., Neems D., Kerschner JL., Kosak ST., Harris A. Architectural proteins CFTR and cohesin have distinct roles in modulating the higher order structure and expression of the CFTR locus Nucleic Acids Research 2014; 42: 9612 - 9622

Guo JH., Chen H., Ruan YC., Zhang XL., Zhang XH., Fok KL., Tsang LL., Yu MK., Huang WQ., Sun X., Chung YW., Jiang XH., Sohma Y., Chan HC. Glucoside-induced electrical activities and insulin secretion in pancreatic islet beta-cells are modulated by CFTR Nature Communications 2014; 5: Arno: 4420

Haddia S., Van Goor F., Zhou JL., Arumugam V., McCartney J., Hazlewood A., Decker C., Negulescu P., Groeninhuys PDJ. Discovery of N-(2,4-Di-tert-butyl-5-hydroxyxypyridine)-4-oxo-1,4-dihydrolquinoline-3- carboxamide (VX-770, Ivacaftor), a Potent and Orally Bioavailable CFTR Potentiator Journal of Medicinal Chemistry 2014; 57: 9776 - 9795


Hill AE., Plyler ZE., Tiwari H., Patki A., Tully JP., McAtee CW., Moseley LA., Sorscher EJ. Longevity and Plasticity of CFTR Provide an Argument for the CFT1 Locus in the Diagnosis of Gastric Cancer Pflugers Journal Of General Physiology 2014; 144: 321 - 336

Lin WY., Jih KY., Hwang TC. A single amino acid substitution in CFTR converts ATP to an inhibitory ligand Journal Of General Physiology 2014; 144: 311 - 320

Linsdell P. State-dependent blocker interactions with the CFTR chloride channel: implications for gating the pore Pflugers Arch-European Journal of Physiology 2014; 466: 2243 - 2255

Liu XH., Dawson DC.
Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Potentiators Protect G551D but Not Triangle F508 CFTR from Thermal Instability
Biochemistry 2014; 53: 5613 - 5618

Mall MA., Harti D.
CFTR: cystic fibrosis and beyond
European Respiratory Journal 2014; 44: 1042 - 1054

Marcourelles P., Friocourt G., Uguen A., Lede F., Ferec C., Laquiere A.
Cystic Fibrosis Transmembrane Conductance Regulator Protein (CFTR) Expression in the Developing Human Brain: Comparative Immunohistochemical Study between Patients with Normal and Mutated CFTR

McShane AJ., Bajrami B., Ramos AA., Diego-Limpi PA., Farrokhi V., Coutemarsh BA., Stanton BA., Jensen T., Riordan JR., Wetmore D., Joesloff E., Yao XD.
Targeted Proteome Quantification of the Absolute Expression and Turnover of Cystic Fibrosis Transmembrane Conductance Regulator in the Apical Plasma Membrane
Journal of Proteome Research 2014; 13: 4676 - 4685

Merkus PJFM.
Setting the stage for CFTR modulator studies in infants
Thorax 2014; 69: 888

Campylobacter jejuni infection suppressed Cl- secretion induced by CFTR activation in T-84 cells
Journal of Infection and Chemotherapy 2014; 20: 682 - 688

Nieddu E., Pollaro B., Mazzei MT., Anzaldi M., Schenehe S., Pedemonte N., Pesce E., Gallietta LJV., Mazzei M.
The search for a common structural moiety among selected pharmacological correctors of the mutant CFTR chloride channel
Future Medicinal Chemistry 2014; 6: 1857 - 1868

Rapid detection of the mature form of cystic fibrosis transmembrane regulator by surface plasmon resonance
Analytical Methods 2015; 7: 226 - 236

Wang W., Roessler BC., Kirk KL.
An Electrostatic Interaction at the Tetrahelix Bundle Promotes Phosphorylation-dependent Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Channel Opening
Journal of Biological Chemistry 2014; 289: 30364 - 30378

Ye L., Hu B., El-Badri F., Hudson BM., Phuan PW., Verkman AS., Tannilo DJ., Kurth MJ.
Delta F508-CFTR correctors: Synthesis and evaluation of thiazole-tethered imidazolines, oxazoles, oxadiazoles, and thiadiazoles
Bioorganic & Medicinal Chemistry Letters 2014; 24: S840 - S844

Zhang XM., Chen QJ., Wang YM., Peng W., Cai H.
Sinupret Activates CFTR and TMEM16A-Dependent Trans epithelial Chloride Transport and Improves Indicators of Mucociliary Clearance
PLoS One 2014; 9: e104970

Zhang S., Skinner D., Hicks SB., Bevensee MO., Sorscher EJ., Lazerak A., Natelson S., McNicholas CM., Woodworth BA.
CFTR potentiators partially restore channel function to A561E CFTR
British Journal of Pharmacology 2014; 171: 4831 - 4848

Clinical
Davies JC., Edbon AM., Orchard C.
Recent advances in the management of cystic fibrosis
Archives of Disease In Childhood 2014; 99: 1033 - 1036

Pseudomonas aeruginosa reduces the expression of CFTR via post-translational modification of NHERF1
Pflügers Arch-European Journal of Physiology 2014; 466: 2269 - 2278

Schutte A., Ermund A., Becker-Pauly C., Johannson MEV., Rodriguez-Pineiro AM., Backhed F., Muller S., Lottadz D., Bond JS.,ansson GC.
Microbial-induced meprin beta cleavage in MUC2 mucin and a functional CFTR channel are required to release anchored small intestinal mucus
Proceedings of the National Academy of Sciences of the United States 2014; 111: 12396 - 12401

Detection of CFTR mutations using PCR/ARMS in a sample of Algerian population
Annales de Biologie Clinique 2014; 72: S49 - S54

Sharma H., Mavaduru RS., Singh SK., Prasad R.
Heterogeneous spectrum of mutations in CFTR gene from Indian patients with congenital absence of the vas deferens and their association with cystic fibrosis genetic modifiers
Molecular Human Reproduction 2014; 20: 827 - 835

Tripathi R., Benz N., Culloton B., Trouve P., Ferec C.
Biophysical Characterisation of Calumenin as a Charged F508del-CFTR Folding Modulator
PLoS One 2014; 9: e104970

Trouve P., Calvez ML., Moisan S., Le Hir S., Huguet F., Benz N., Kerbiriou M., Ferec C.
Rapid detection of the mature form of cystic fibrosis transmembrane regulator by surface plasmon resonance
Analystical Methods 2015; 7: 226 - 236

Ye L., Hu B., El-Badri F., Hudson BM., Phuan PW., Verkman AS., Tannilo DJ., Kurth MJ.
Delta F508-CFTR correctors: Synthesis and evaluation of thiazole-tethered imidazolines, oxazoles, oxadiazoles, and thiadiazoles
Bioorganic & Medicinal Chemistry Letters 2014; 24: S840 - S844

Zhang XM., Chen QJ., Wang YM., Peng W., Cai H.
Sinupret Activates CFTR and TMEM16A-Dependent Trans epithelial Chloride Transport and Improves Indicators of Mucociliary Clearance
PLoS One 2014; 9: e104970

Zhang S., Skinner D., Hicks SB., Bevensee MO., Sorscher EJ., Lazerak A., Natelson S., McNicholas CM., Woodworth BA.
CFTR potentiators partially restore channel function to A561E-CFTR, a cystic fibrosis mutant with a similar mechanism of dysfunction as F508del-CFTR
British Journal of Pharmacology 2014; 171: 4490 - 4503
De Boeck K., Castellani C., Elborn JS.
Medical consensus, guidelines, and position papers: A policy for the ECFS
Journal of Cystic Fibrosis 2014; 13: 495 - 498

Doul I.
Cystic Fibrosis Papers of the Year 2013
Paediatric Respiratory Reviews 2014; 15: 10 - 12

Ezzati A., Batoei F., Jafari SA., Kiyani MA., Mandavi-Shahri N., Ahanchian H., Tehrani S., Kianifar HR.
Dermatoglyphic Patterns in Cystic Fibrosis Children
Iranian Journal Of Pediatrics 2014; 24: 609 - 616

Hubert D., Soubeiran L., Gourmelon F., Grenet D., Serreau R., Perrodeau E., Zegarra-Parodi R., Bposts T.
Impact of Osteopathic Treatment on Pain in Adult Patients with Cystic Fibrosis - A Pilot Randomized Controlled Study
PloS One 2014; 9: 7:e102946

Jennings MT., Riekert KA., Boyle MP.
Update on Key Emerging Challenges in Cystic Fibrosis
Medical Principles and Practice 2014; 23: 393 - 402

Katz ES.
Cystic Fibrosis and Sleep
Clinics In Chest Medicine 2014; 35: 495

Kintu B., Brightwell A.
Episodic Seasonal Pseudo-Bartter Syndrome in Cystic Fibrosis
Paediatric Respiratory Reviews 2014; 15: 19 - 21

Liu LC., Shyr SD., Chu SH., Huang LH., Kao YH., Lei WT., Cheng CH., Lo CY., Chen CK., Fang LC.
Cystic fibrosis: Experience in one institution
Journal of Microbiology Immunology and Infection 2014; 47: 358 - 361

MacKenzie T., Gifford AH., Sabadosa KA., Quinton HB., Knapp EA., Goss CH., Marshall BC.
Longevity of Patients With Cystic Fibrosis in 2000 to 2010 and Beyond: Survival Analysis of the Cystic Fibrosis Foundation Patient Registry
Annals of Internal Medicine 2014; 161: 233

The development and deployment of integrated electronic care records in a regional adult and paediatric cystic fibrosis unit

Pilewski JM., Taichman DB.
Cystic Fibrosis: Recent Successes Present New Challenges
Annals of Internal Medicine 2014; 161: 298

Retsch-Bogart GZ., Van Dalens JM., Marshall BC., George C., Pilewski JM., Nelson EC., Goss CH., Ramsey BW.
Highly Effective Cystic Fibrosis Clinical Research Teams: Critical Success Factors
Journal of General Internal Medicine 2014; 29:

Silla IO., Pueyo JIM., Aroz SG., Vigo FD.
Amyloid goiter in a patient with cystic fibrosis
Medicina Clinica 2014; 143: 517 - 518

The value of soluble transferrin receptor and hepcidin in the assessment of iron status in children with cystic fibrosis

Walshaw M.
Highlights of the North American CF Conference 2013
Paediatric Respiratory Reviews 2014; 15: 8 - 9

[Anonymous].
Supplement: The 28th Annual North American Cystic Fibrosis Conference Georgia World Congress Center, Atlanta, Georgia, October 9-11, 2014 Abstracts
Pediatric Pulmonology 2014; 49:

Diabetes

Coriati A., Belson L., Ziai S., Haberer E., Gauthier MS., Mailhot G., Coderre L., Berthiaume Y., Rabasa-Lhoret R.
Impact of Sex on Insulin Secretion in Cystic Fibrosis
Journal of Clinical Endocrinology & Metabolism 2014; 99: 1767 - 1773

Delaney RA., Windemuth B.
The Unique Management of Cystic Fibrosis-Related Diabetes and the Importance of Glycemic Control

Hayes D., McCoy KS., Sheikh SI.
Resolution of Cystic Fibrosis-related Diabetes with Ivacaftor Therapy
American Journal of Respiratory and Critical Care Medicine 2014; 190: 590 - 591

Middleton PG., Matson AG., Robinson PD., Holmes-Walker DJ., Katz T., Hameed S.
Cystic Fibrosis Related Diabetes: Potential pitfalls in the transition from paediatric to adult care
Paediatric Respiratory Reviews 2014; 15: 281 - 284

Moran A., Pillay K., Becker DJ., Acerni CL.
Management of cystic fibrosis-related diabetes in children and adolescents
Pediatric Diabetes 2014; 15: 65 - 76

O'Shea D., O'Connell J.
Cystic Fibrosis Related Diabetes
Current Diabetes Reports 2014; 14:

Perano S., Rayner CK., Couper J., Martin J., Horowitz M.
Cystic fibrosis related diabetes-a new perspective on the optimal management of postprandial glycaemia
Journal of Diabetes and its Complications 2014; 28: 904 - 911

Rayas MS., Willey-Courand DB., Lynch JL., Guajardo JR.
Improved Screening for Cystic Fibrosis-Related Diabetes by an Integrated Care Team Using an Algorithm
Pediatric Pulmonology 2014; 49: 971 - 977

Scheuing N., Berger G., Bergis D., Gohlke B., Konrad K., Laubner K., Lilienthal E., Moser C., Schutz-Fuhrmann I., Thon A., Holl RW.
Adherence to clinical care guidelines for cystic fibrosis-related diabetes in 659 German/Austrian patients
Journal of Cystic Fibrosis 2014; 13: 730 - 736

Smerieri A., Montanini L., Maiuri L., Bernasconi S., Street MM.
FOXO1 Content Is Reduced in Cystic Fibrosis and Increases with IGF-1 Treatment
International Journal of Molecular Sciences 2014; 15: 18000 - 18022

Wickens-Mitchell KL., Gilchrist FJ., McKenna D., Raffeeq P., Lenney W.
The screening and diagnosis of cystic fibrosis-related diabetes in the United Kingdom

Diagnosis

Bagheri-Hanso A., Nedwed S., Rueckes-Nilges C., Naehrlich L.
Intestinal current measurement versus nasal potential difference measurements for diagnosis of cystic fibrosis: a case-control study
BMC Pulmonary Medicine 2014; 14: ArNo: 156

D’Erme AM., Braggion C., de Martino M., Greco A.
Aquagenic palmoplantar keratoderma: a sign of cystic fibrosis early in life
International Journal of Dermatology 2014; 53: E536 - E538
Hill M., Compton C., Karunaratna M., Lewis C., Chitty L. 
Client Views and Attitudes to Non-Invasive Prenatal Diagnosis for Sickle Cell Disease, Thalassaemia and Cystic Fibrosis. Journal of Genetic Counseling 2014; 23: 1012 - 1021

Kuban P., Gregus M., Pokojova E., Skrickova J., Foret F. 
Double opposite end injection capillary electrophoresis with contactless conductometric detection for simultaneous determination of chloride, sodium and potassium in cystic fibrosis diagnosis. Journal of Chromatography A 2014; 1358: 293 - 298

Matta ACV., Leone C., Rodrigues JC., Adve FF. 

Label-Free Efficient and Accurate Detection of Cystic Fibrosis Causing Mutations using an Azimuthally Rotated GC-SPR Platform Analytical Chemistry 2014; 86: 11773 - 11781

Poulou M., Destouni A., Kakourou G., Kanavakis E., Tzetis M. 

Rock MJ., Makholm L., Eickhoff J. 

Welsh SK., Gross JE., Larson NS., Berg JM., Roy D., Pinker JE. 

Epidemiology

Ivady G., Koczok K., Madar L., Gombos E., Toth I., Gyori K., Balogh I. 
Molecular analysis of cystic fibrosis patients in hungary - an update to the mutational spectrum using an Azimuthally Rotated GC-SPR Platform Analytical Chemistry 2014; 34: 46 - 51

Stojanovic KS., Hubert D., Leroy S., Dominique S., Grenet D., Colombat M., Clement A., Fayon M., Grateau G. 

Exercise

Cohen SP., Orenstein DM. 

Collaco JM., Blackman SM., Raraigh KS., Morrow CB., Cutting GR., Paranjape SM. 
Self-reported exercise and longitudinal outcomes in cystic fibrosis: a retrospective cohort study. BMC Pulmonary Medicine 2014; 14: ArNo: 159

Cox NS., Alison JA., Holland AE. 
Interventions to promote physical activity in people with cystic fibrosis. Paediatric Respiratory Reviews 2014; 15: 237 - 239

Franco CB., Ribeiro AF., Vlorcillo AM., Zambon MP., Almeida MB., Rozov T. 
Effects of Pilates mat exercises on muscle strength and on pulmonary function in patients with cystic fibrosis. Jornal Brasileiro de Pneumologia 2014; 40: 521 - 527

Gastroenterology

Billing JS., Dunitz JM., McAllister S., Herzog T., Bobr A., Khoruts A. 


Gory I., Brown G., Wilson J., Kemp W., Paul E., Roberts SK. 
Increased risk of colorectal neoplasia in adult patients with cystic fibrosis: a matched case-control study. Scandinavian Journal of Gastroenterology 2014; 49: 1230 - 1236

Ledder O., Haller W., Coupe RTL., Lewindon P., Oliver M. 

Li L., Somerset S. 
Digestive system dysfunction in cystic fibrosis: Challenges for nutrition therapy. Digestive and Liver Disease 2014; 46: 865 - 874

Perano SJ., Couper JJ., Horowitz M., Martin AJ., Kritas S., Sullivan T., Rayner CK. 
Pancreatic Enzyme Supplementation Improves the Incretin Hormone Response and Attenuates Postprandial Glycemia in Adolescents With Cystic Fibrosis: A Randomized Crossover Trial. Journal of Clinical Endocrinology & Metabolism 2014; 99: 2486 - 2493

Roberts K., Liu I., Jaffe A., Verge CF., Thomas PS. 

Seegeimiller AC. 
Subhi R., Ooi R., Finlayson F., Kotsimbos T., Wilson J., Lee WR., Wale R., Warrier S.  
ANZ Journal Of Surgery 2014; 84: 740 - 744

Prediction of acute pancreatitis risk based on PIP score in children with cystic fibrosis  
Journal of Cystic Fibrosis 2014; 13: 579 - 584

Trang T., Chan J., Graham DY.  
Pancreatic enzyme replacement therapy for pancreatic exocrine insufficiency in the 21st century  
World Journal of Gastroenterology 2014; 20: 11467 - 11485

Wooldridge JL., Schaeffer D., Jacobs D., Thieroff Ekerdt R.  
Long-Term Experience With ZENPEP in Infants With Exocrine Pancreatic Insufficiency Associated With Cystic Fibrosis  
Journal Of Pediatric Gastroenterology And Nutrition 2014; 59: 612 - 615

Gene Therapy

Davies LA., Nunez-Alonso GA., McLachlan G., Hyde SC., Gill DR.  
Aerosol Delivery of DNA/Liposomes to the Lung for Cystic Fibrosis  
Gene Therapy  

Gill DR., Hyde SC.  
Delivery of genes into the CF airway  
Thorax 2014; 69: 962 – 964

Genetics

Anderson RL., Murray K., Chong JX., Owenga R., Antillon M., Chen P., de Leon LD., Swoboda KJ., Lester LA., Das S., Ober C., Waggoner DJ.  
Disclosure of Genetic Research Results to Members of a Founder Population  
Journal of Genetic Counseling 2014; 23: 984 - 991

Cutting GR.  
Cystic fibrosis genetics: from molecular understanding to clinical application  
Nature Reviews Genetics 2015; 16: 45 - 56

Ferril GR., Nick JA., Gotz AE., Barham HP., Saavedra MT., Taylor-Cousar JL., Nichols DP., Curran-Everett D., Kingdom TT., Ramakrishnan VR.  
Comparison of radiographic and clinical characteristics of low-risk and high-risk cystic fibrosis genotypes  
International Forum of Allergy & Rhinology 2014; 4: 915 - 920

Guo X., Pace RG., Stoneraker JR., O’Neal WK., Knowles MR.  
Meconium ileus in cystic fibrosis is not linked to central repetitive region length variation in MUC1, MUC2, and MUC5AC  
Journal of Cystic Fibrosis 2014; 13: 613 - 616

Lenarduzzi S., Morgutti M., Crovella S., Coiana A., Rosatelli MC.  
Novel truncating mutations in the CFTR gene causing a severe form of cystic fibrosis in Italian patients  
Genetics and Molecular Research 2014; 13: 9636 - 9641

Melotti P., Mafficini A., Lebecque P., Ortombina M., Leal T., Pintani E., Pepermans X., Sorio C., Assael BM.  
Impact of MIF Gene Promoter Polymorphism on F508del Cystic Fibrosis Patients  
PloS One 2014; 9: 12:e114274

Growth & Development

Hanna RM., Weiner DJ.  
Overweight and Obesity in Patients With Cystic Fibrosis: A Center-Based Analysis  
Pediatric Pulmonology 2015; 50: 35 - 41

Heitse SL., Borowitz DS., Leung DH., Ramsey B., Mayer-Hamblett N.  
Early attained weight and length predict growth faltering better than velocity measures in infants with CF  
Journal of Cystic Fibrosis 2014; 13: 723 - 729

Compromised Bone Microarchitecture and Estimated Bone Strength in Young Adults With Cystic Fibrosis  
Journal of Clinical Endocrinology & Metabolism 2014; 99: 3399 – 3407

Immunology & Inflammation

Bezzerril V., Avitabile C., Decheccchi MC., Lampronti I., Borgatti M., Montagner G., Cabrini G., Gambari R., Romanelli A.  
Antibacterial and anti-inflammatory activity of a temporin B peptide analogue on an in vitro model of cystic fibrosis  
Journal of Peptide Science 2014; 20: 822 - 830

Boikos C., De Serres G., Lands LC., Boucher FD., Tapiero B., Dainegault P., Quach C.  
Safety of Live-Attenuated Influenza Vaccination in Cystic Fibrosis  
Pediatrics 2014; 134: E983 - E991

Browning MJ., Lim MTC., Kenia P., Whittle M., Doftinger R., Barcenas-Morales G., Kumararatne D., Viskaduraki M., O’Callaghan C., Gaillard EA.  
Pneumococcal polysaccharide vaccine responses are impaired in a subgroup of children with cystic fibrosis  
Journal of Cystic Fibrosis 2014; 13: 632 - 638

de Vries L., Griffiths A., Armstrong D., Robinson PJ.  
Cytokine gene polymorphisms and severity of CF lung disease  

Dwyer M., Shan Q., D’Ortona S., Maurer R., Mitchell R., Olesen H., Thiel S., Hubeiner J., Gadjeva M.  
Cystic Fibrosis Sputum DNA Has NETHosis Characteristics and Neutrophil Extracellular Trap Release Is Regulated by Macrophage Migration-Inhibitory Factor  
Journal of Innate Immunity 2014; 6: 765 - 779

Fischer N., Hentschel J., Markert UR., Keller PM., Pletz MW., Mainz JG.  
Non-Invasive Assessment of Upper and Lower Airway Infection and Inflammation in CF Patients  
Pediatric Pulmonology 2014; 49: 1065 - 1075

Grassme H., Carpinetto A., Edwards MJ., Gulbins E., Becker KA.  
Regulation of the Inflammasome by Ceramide in Cystic Fibrosis  
Lungs  
Cellular Physiology and Biochemistry 2014; 34: 45 - 55

Hofer TP., Frankenberger M., Heimbeck I., Burggraf D., Wjst M., Wright AKA., Kerscher M., Nahrig S., Huber RM., Fischer R., Helbtorr L.  
Decreased expression of HLA-DQ and HLA-DR on cells of the monocytic lineage in cystic fibrosis  
Journal of Molecular Medicine 2014; 92: 1293 - 1304
Kappler M., Nagel F., Feilcke M., Heilig G., Grimmelt AC., Pawlita I., Irnstetter A., Hildebrandt J., Burmester H., Kroner C., Griese M. 
Predictive values of antibodies against Pseudomonas aeruginosa in patients with cystic fibrosis one year after early eradication treatment 
Journal of Cystic Fibrosis 2014; 13: 534 - 541

Kragh KN., Alhede M., Jensen PO., Moser C., Scheike T., Jacobsen CS., Poulsen SS., Eckhardt-Sorensen SR., Trostrup H., Christoffersen L., Hougen HP., Rickelt LF., Kuhl M., Hoiby N., Bjarnsholt T. 
Polymorphonuclear Leukocytes Restrict Growth of Pseudomonas aeruginosa in the Lungs of Cystic Fibrosis Patients 
Infection and Immunity 2014; 82: 4477 - 4486

Mauch RM., Levy CE. 
Serum antibodies to Pseudomonas aeruginosa in cystic fibrosis as a diagnostic tool: A systematic review 

Mauch RM., Rossi CL., Ribeiro JD., Ribeiro AF., da Silva MN., Levy CE. 
Assessment of IgG antibodies to Pseudomonas aeruginosa in patients with cystic fibrosis by an enzyme-linked immunosorbent assay (ELISA) 
Diagnostic Pathology 2014; 9: ArNo: 158

McElvaney OJ., O’Reilly N., White M., Lacey N., Pohl K., Gerzta T., Bergin DA., Kerr H., McCarthy C., O’Brien ME., Adage T., Kungl AJ., Reeves EP., McElvaney NG. 
The effect of the decoy molecule PA401 on CXCL8 levels in patients with cystic fibrosis 
Molecular Immunology 2015; 63: 550 - 558

Morrow CB., Raleigh KS., Green DM., Blackman SM., Cutting GR., Collaco JM. 
Cat and Dog Exposure and Respiratory Morbidities in Cystic Fibrosis 
Journal of Pediatrics 2014; 165: 830 - 842

Musson JA., Reynolds CJ., Rinchai D., Nithichanon A., Khaenam P., Favry E., Spink N., Chu KKY., De Soyza A., Reynolds OJ., O'Reilly T., Altmann DM., Robinson JH. 
CD4(+) T Cell Epitopes of FliC Conserved between Strains of P. aeruginosa 
Molecular Immunology 2015; 63: 501 - 507

Pabary R. 
Severe pulmonary exacerbation in cystic fibrosis caused by cat allergy 
Paediatric Respiratory Reviews 2014; 15: 29 - 31

Palomo J., Marcich T., Pirotet J., Fauconnier L., Robinet M., Reverchon F., Le Bert M., Topge D., Buijs Offerman R., Stolarczyk M., Quesniaux VFJ., Scholte BJ., Ryffel B. 
Role of IL-1 beta in Experimental Cystic Fibrosis upon P. aeruginosa Infection 
PloS One 2014; 9: e14884

Peetermans M., Goeminne P., De Boeck C., Dupont LJ. 
IgE Sensitization to Aspergillus fumigatus Is Not a Bystander Phenomenon in Cystic Fibrosis Lung Disease 
Chest 2014; 144: E99 - E100

Sahli C., Fredj SH., Siala H., Bibi A., Messaoud T. 
First study of angiotensin converting enzyme in cystic fibrosis Tunisian patients 
Clinical Chemistry and Laboratory Medicine 2014; 59: E211 - E215

Neutrophil elastase-mediated increase in airway temperature during inflammation 

Invariant Natural Killer T (iNKT) Cells Prevent Autoimmunity, but Induce Pulmonary Inflammation in Cystic Fibrosis 
Cellular Physiology and Biochemistry 2014; 34: 56 - 70

Differential expression of IL-33 and HMGB1 in the lungs of stable cystic fibrosis patients 
European Respiratory Journal 2014; 44: 802 - 805

Uriarte SM. 
Novel insights related to CF neutrophils 
Blood 2014; 124: 985 - 986

Zicari AM., Celani C., De Castro G., De Biase RV., Duse M. 
Anti IgE antibody as treatment of allergic bronchopulmonary aspergillosis in a patient with cystic fibrosis 
European Review for Medical and Pharmacological Sciences 2014; 18: 1839 – 1841

Liver Disease

Martin FM., Malagon AIM., Gonzalez IJA., Luis HD., Pallares ALC. 
Hepatolithiasis in cystic fibrosis: A special condition for surgical treatment 
Cirugia Espanola 2014; 92: 634 - 635

Staufer K., Hallibasic E., Trauner M., Kazemi-Shirazi L. 
Cystic Fibrosis Related Liver Disease—Another Black Box in Hepatology 
International Journal of Molecular Sciences 2014; 15: 13529 - 13549

Wagener JS., Woo MS., Pasta DJ., Konstan MW., Morgan WJ. 
Liver Involvement in the Hispanic Population of North America With Cystic Fibrosis 
Journal Of Pediatric Gastroenterology And Nutrition 2014; 59: 476 – 479

Microbiology

Adjemian J., Olivier KN., Prevots DR. 
Nontuberculous Mycobacteria among Patients with Cystic Fibrosis in the United States Screening Practices and Environmental Risk 
American Journal of Respiratory and Critical Care Medicine 2014; 190: 581 - 586

Ahmed B., Bush A., Davies JC. 
How to use: bacterial cultures in diagnosing lower respiratory tract infections in cystic fibrosis 
Mycobacterium abscessus complex
Advances in the management of pulmonary disease due to SE.
Kalampouka E., Petrocheilou A., Kaditis AG., Doudounakis
Journal of Innate Immunity 2014; 6: 846

Jovic S., Shikhagae M., Morgelin M., Kjellstrom S., Erjefalt J., Ling KM., Looi K., Kicic A.
Determinants of culture success in an airway epithelium sampling program of young children with cystic fibrosis
Experimental Lung Research 2014; 40: 447 - 459

Guio A., Buendia B., Llorca L., Punter RMG., Giron R.
Chrysobacterium spp., a new opportunistic pathogen associated with cystic fibrosis?
Enfermedades Infecciosas Y Microbiologia Clinica 2014; 32: 497 - 501

Harris KA., Kenna DTD.
Mycobacterium abscessus infection in cystic fibrosis: molecular typing and clinical outcomes
Journal of Medical Microbiology 2014; 63: 1241 - 1246

Hosseinkhan N., Zarrineh P., Masoudi Nejad A.
Analysis of Genome-scale Expression Network in Four Major Bacterial Residents of Cystic Fibrosis Lung
Current Genomics 2014; 15: 408 - 418

Jackson AA., Daniels EF., Hammond JH., Willger SD., Hogan DA.
Global regulator Arr represses PlcH phospholipase activity in Pseudomonas aeruginosa when oxygen is limiting
Microbiology-SGM 2014; 160: 2215 - 2225

Jeon SM., Lim NR., Kwon SJ., Shim TS., Park MS., Kim BJ., Kim SH.
Analysis of species and intra-host diversification of Pseudomonas aeruginosa using pulsed-field gel electrophoresis (PFGE) and multi-locus sequence typing (MLST)
Journal of Microbiological Methods 2014; 103: 19 - 25

Jovic S., Shikhagae M., Morgelin M., Kjellstrom S., Erjefalt J., Olin AI., Frick IM., Egesten A.
Expression of MIG/CXCL9 in Cystic Fibrosis and Modulation of Its Activities by Elastase of Pseudomonas aeruginosa
Journal of Innate Immunity 2014; 6: 846 - 859

Kalampouka E., Petrocheilou A., Kaditis AG., Doudounakis SE.
SI45SX CFTR Mutation and Upper Airway Colonization With Pseudomonas aeruginosa
Archivos de Bronconeumologia 2014; 50: 499 - 500

Koh WJ., Stout JE., Yew WW.
Advances in the management of pulmonary disease due to Mycobacterium abscessus complex
International Journal of Tuberculosis and Lung Disease 2014; 18: 1141 - 1149

Line L., Alhede M., Kolpen M., Kuhl M., Ciofu O., Bjarnsholt T., Moser C., Toyofuku M., Nomura N., Holby N., Jensen PO.
Physiological levels of nitrate support anoxic growth by denitrification of Pseudomonas aeruginosa at growth rates reported in cystic fibrosis lungs and sputum
Frontiers In Microbiology 2014; 5: ArNo: 554

Mehenthaliringam E.
Emerging cystic fibrosis pathogens and the microbiome
Paediatric Respiratory Reviews 2014; 15: 13 - 15

Marguerettaz M., Dieppois G., Que YA., Ducret V., Zuchuat S., Perron K.
Sputum containing zinc enhances carbapenem resistance, biofilm formation and virulence of Pseudomonas aeruginosa
Microbial Pathogenesis 2014; 77: 36 - 41

Markussen T., Marvig RL., Gomez-Lozano M., Aanaes K., Burleigh AE., Hoiby N., Johansen HK., Molin S., Jelsbak L.
Environmental Heterogeneity Drives Within-Host Diversification and Evolution of Pseudomonas aeruginosa
MBio 2014; 5: S:e01592-14

Pseudomonas aeruginosa Phenotypes Associated With Eradication Failure in Children With Cystic Fibrosis
Clinical Infectious Diseases 2014; 59: 624 - 631

McAdam PR., Richardson EJ., Fitzgerald JR.
High-throughput sequencing for the study of bacterial pathogen biology
Current Opinion In Microbiology 2014; 19: 106 - 113

Menezes FG., Abreu MGB., Kawagoe JY., Warth AN., Deutsch AD., Dornaus MFPS., Martino MD., Correa L.
Ochrobactrum anthropi bacteremia in a preterm infant with cystic fibrosis
Brazilian Journal of Microbiology 2014; 45: 559 - 561

Michalska K., Chhor G., Joachimiak A.
RsaM: a transcriptional regulator of Burkholderia spp. with novel fold
FEBS Letters 2014; 581: 14 - 22

Min KB., Lee KM., Oh YT., Yoon SS.
Nonmucoid conversion of mucoid Pseudomonas aeruginosa induced by sulfate-stimulated growth
FEBS Microbiology Letters 2014; 360: 157 - 166

Moore JE., Rao JR.
Efficacy of the biocide Steri-7 against the common Gram-negative bacterial pathogens (Burkholderia cenocepacia, Burkholderia gladioli, Burkholderia multivorans, Pseudomonas aeruginosa and Stenotrophomonas maltophilia) associated with cystic fibrosis
British Journal of Biomedical Science 2013; 70: 80 - 81

Moore JE., Rendall JC.
Comparison of susceptibility of cystic-fibrosis-related and non-cystic-fibrosis-related Pseudomonas aeruginosa to chlorine-based disinfecting solutions: implications for infection prevention and ward disinfection
Journal of Medical Microbiology 2014; 63: 1214 - 1219

Moscoso JA., Jaeger T., Valentini M., Hui KL., Jenal U., Filoux A.
The Diguanylate Cyclase SdcC Is a Central Player in Gac/Rsm-Mediated Biofilm Formation in Pseudomonas aeruginosa
Journal of Bacteriology 2014; 196: 4081 - 4088

Nielsen SM., Kristensen L., Sondergaard A., Handberg KJ., Stenderup J., Norskov-Lauritsen N.
Increased prevalence and altered species composition of filamentous fungi in respiratory specimens from cystic fibrosis patients
APMIS 2014; 122: 1007 - 1012

Inhaled corticosteroids and Aspergillus fumigatus isolation in cystic fibrosis
Medicinal Mycology 2014; 52: 712 - 722


Impact of Scedosporium apiospermum complex seroprevalence in patients with cystic fibrosis
Journal of Cystic Fibrosis 2014; 13: 667 - 673

Pashley CH.

Fungal Culture and Sensitisation in Asthma, Cystic Fibrosis and Chronic Obstructive Pulmonary Disorder: What Does It Tell Us?
MycoPathologia 2014; 178: 457 - 463


Sphingoid long chain bases prevent lung infection by Pseudomonas aeruginosa
EMBO Molecular Medicine 2014; 6: 1205 - 1214

Poole K.

Stress responses as determinants of antimicrobial resistance in Pseudomonas aeruginosa: multidrug efflux and more
Canadian Journal of Microbiology 2014; 60: 783 - 791

Prityktova T., Lighty TJ., Kumar B., Bernier SP., Sorenson JL., Surette MG., Cardona ST.

The attenuated virulence of a Burkholderia cenocepacia paaABCD mutant is due to inhibition of quorum sensing by release of phenylacetic acid
Molecular Microbiology 2014; 94: 522 - 536

Qvist T., Johansen IS., Pressler T., Holby N., Andersen AB., Katzenstein TL., Bjerrum S.

Urinary lipoarabinomannan point-of-care testing in patients affected by pulmonary nontuberculous mycobacteria - experiences from the Danish Cystic Fibrosis cohort study
BMC Infectious Diseases 2014; 14: ArNo: 655

Sahoo M., del Barrio L., Miller MA., Re F.

Neutrophil Elastase Causes Tissue Damage That Exposes the Epithelial Barrier to Pseudomonas aeruginosa Biofilm Pump Activity
PLoS One 2014; 9: 8:e109896

Satana D., Erkose-Genc G., Tamay Z., Uzun M., Guler N., Erturan Z.

Prevalence and drug resistance of mycobacteria in Turkish cystic fibrosis patients
Annals of Clinical Microbiology and Antimicrobials 2014; 13: ArNo: 28

Savoia D.

New perspectives in the management of Pseudomonas aeruginosa infections
Future Microbiology 2014; 9: 917 - 928

Schwab U., Abdullah LH., Perlmutt OS., Albert D., Davis CW., Arnold RR., Yankaskas JR., Gilligan P., Neubauer H., Randell SH., Boucher RC.

Localization of Burkholderia cepacia Complex Bacteria in Cystic Fibrosis Lungs and Interactions with Pseudomonas aeruginosa in Hypoxic Mucus
Infection and Immunity 2014; 82: 4729 - 4745

Sherrard LJ., Schable B., Graham KA., McGrath SJ., McIlreavey L., Hatch J., Wolfgang MC., Muhlebach MS., Gilpin DF., Schneiders T., Elborn JS., Tunney MM.

Mechanisms of reduced susceptibility and genotypic prediction of antibiotic resistance in Pseudomonas aeruginosa isolates from cystic fibrosis (CF) and non-CF patients
Journal of Antimicrobial Chemotherapy 2014; 69: 2690 - 2698

Sherrard LJ., Tunney MM., Elborn JS.

Antimicrobial resistance in the respiratory microbiota of people with cystic fibrosis
Lancet 2014; 384: 703 - 713

Short FL., Murdoch SL., Ryan RP.

Polybacterial human disease: the ill of social networking
Trends In Microbiology 2014; 22: 508 - 516

Smith DJ., Badrick AC., Zakrzewski M., Krause L., Bell SC., Anderson GJ., Reid DW.

Pyrosequencing reveals transient cystic fibrosis lung microbiome changes with intravenous antibiotics
European Respiratory Journal 2014; 44: 922 - 930

Smith DJ., Hill GR., Bell SC., Reid DW.

Reduced Mucosal Associated Invariant T-Cells Are Associated with Increased Disease Severity and Pseudomonas aeruginosa Infection in Cystic Fibrosis


Identification of Anti-virulence Compounds That Disrupt Quorum-Sensing Regulated Acute and Persistent Pathogenicity
PLoS Pathogens 2014; 10: 8:e1004321

Stokell JR., Gharibeh RZ., Hamp TJ., Zapata MJ., Fodor AA., Steck TR.

Analysis of Changes in Diversity and Abundance of the Microbial Community in a Cystic Fibrosis Patient over a Multiyear Period

Tolker-Nielsen T.

Pseudomonas aeruginosa biofilm infections: From molecular biofilm biology to new treatment possibilities
APMIS 2014; 122: 1-51

Torres IMS., Patankar YR., Shabanee TB., Dolben E., Hogan DA., Leib DA., Berwin BL.

Acidosis Potentiates the Host Proinflammatory Interleukin-1beta Response to Pseudomonas aeruginosa Infection
Infection and Immunity 2014; 82: 4689 - 4697

Tran CS., Rangel SM., Almblad H., Kierbel A., Givskov M., Tolker-Nielsen T., Hauser AR., Engel JN.

The Pseudomonas aeruginosa Type III Translocin Is Required for Biofilm Formation at the Epithelial Barrier

Tseng SP., Tsai WC., Liang CY., Lin YS., Huang JW., Chang CY., Tyan YC., Lu PL.

PLoS One 2014; 9: 8:e104986

Wahab AA., Taj-Aldeen SJ., Kolecia A., ElGindi M., Finkel JS., Boekhout T.

High prevalence of Candida dubliniensis in lower respiratory tract secretions from cystic fibrosis patients may be related to increased adherence properties
International Journal Of Infectious Diseases 2014; 24: 14 - 19

Warris A.

The biology of pulmonary aspergillus infections
Journal of Infection 2014; 69: S36 - S51

Wilkins M., Hall-Stoodley L., Allan RN., Faust SN.

New approaches to the treatment of biofilm-related infections
Journal of Infection 2014; 69: S51 - S52

Wu XM., Chen J., Li XB., Zhao YP., Zughayer SM.

Culture-free diagnostics of Pseudomonas aeruginosa infection by silver nanorod array based SERS from clinical sputum samples
Nanomedicine-Nanotechnology Biology and Medicine 2014; 10: 1863 - 1870
Yahr T., Wozniak D.
The Pseudomonas aeruginosa AlgZR two-component system coordinates multiple phenotypes
Frontiers In Cellular and Infection Microbiology 2014; 4: ArNo: 82

Zemanick ET., Emerson J., Thompson V., McNamara S., Morgan W., Gibson RL., Rosenfeld M.
Clinical Outcomes After Initial Pseudomonas Acquisition in Cystic Fibrosis
Pediatric Pulmonology 2015; 50: 42 - 48

Zhang YN., Shao XL., Wang SH., Zhang SQ.
Effect of Glutathione on Pyocyanin Production in Pseudomonas aeruginosa
Asian Journal of Chemistry 2014; 26: 3265 - 3269

Zlosnik JEA., Mori PY., To D., Leung J., Hird TJ., Speert DP.
Swimming Motility in a Longitudinal Collection of Clinical Isolates of Burkholderia cepacia Complex Bacteria from People with Cystic Fibrosis
Plos One 2014; 9: 9:e106428

Nutrition
Effect of supplementary zinc on body mass index, pulmonary function and hospitalization in children with cystic fibrosis
Turkish Journal of Pediatrics 2014; 56: 127 - 132

Groeleau V., Schall JJ., Dougherty KA., Latham NE., Maqbool A., Mascarenhas MR., Stallings VA.
Effect of a dietary intervention on growth and energy expenditure in children with cystic fibrosis
Journal of Cystic Fibrosis 2014; 13: 572 - 578

Marcondes NA., Raimundo FV., Vanacor R., Corte BP., Ascoli AM., de Azambuja AZ., Scopec L., dos Santos PV., Dalci PDR., Rotta LN., Furlanetto TW., Faulhaber GAM.
Hypovitaminosis D in patients with cystic fibrosis: a cross-section study in South Brazil
Clinical Respiratory Journal 2014; 8: 455 - 459

Siwamogsatham O., Dong W., Binongo JN., Chowdhury R., Alvarez JA., Feinman SJ., Enders J., Tangpricha V.
Relationship Between Fat-Soluble Vitamin Supplementation and Blood Concentrations in Adolescent and Adult Patients With Cystic Fibrosis
Nutrition In Clinical Practice 2014; 29: 491 - 497

Umlawska W., Kryzanowska M., Zielinska A., Sands D.
Effect of Selected Factors Associated with the Clinical Course of the Disease on Nutritional Status in Children with Cystic Fibrosis
Advances In Clinical and Experimental Medicine 2014; 23: 775 – 783

Physiotherapy
Mcllwaine MP., Son NML., Richmond ML.
Physiotherapy and cystic fibrosis: what is the evidence base?
Current Opinion In Pulmonary Medicine 2014; 20: 613 – 617

Psychosocial
Balfour L., Armstrong M., Holly C., Gaudet E., Aaron S., Tasca G., Cameron W., Pakhale S.
Development and psychometric validation of a cystic fibrosis knowledge scale
Respirology 2014; 19: 1209 - 1214

Balfour-Lynn IM.
Personalised medicine in cystic fibrosis is unaffordable
Paediatric Respiratory Reviews 2014; 15: 2 - 5

Bilton D.
Personalised medicine in cystic fibrosis must be made affordable
Paediatric Respiratory Reviews 2014; 15: 6 - 7

del Corral T., Percegona J., Seborga M., Rabinovich RA., Vílaro J.
Physiological response during activity programs using Wii-based video games in patients with cystic fibrosis (CF)
Journal of Cystic Fibrosis 2014; 13: 706 - 711

dos Santos DMDs., Deon KC., Bullinger M., dos Santos CB.
Validity of the DISABKIDS (R) - Cystic Fibrosis Module for Brazilian children and adolescents

Duff AJA., Abbott J., Cowperthwaite C., Sumner C., Hurley MA., Quittner A.
Depression and anxiety in adolescents and adults with cystic fibrosis in the UK: A cross-sectional study
Journal of Cystic Fibrosis 2014; 13: 745 - 753

Duff AJA., Latchford G.
Adherence in cystic fibrosis; care teams need to change first
Lancet Respiratory Medicine 2014; 2: 683 - 685

Health related quality of life and disease burden of patients with cystic fibrosis and their caregivers: Results of the European BURQOL- RD survey in Hungary
Orvosi Hetilap 2014; 155: 1673 - 1684

Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers: results of The International Depression Epidemiological Study
across nine countries
Thorax 2014; 69: 1090 - 1097

Snell C., Fernandes S., Bujoreanu IS., Garcia G.
Depression, Illness Severity, and Healthcare Utilization in Cystic Fibrosis
Pediatric Pulmonology 2014; 49: 1177 - 1181

Wildman MJ., Hoo ZH.
Moving cystic fibrosis care from rescue to prevention by embedding adherence measurement in routine care
Paediatric Respiratory Reviews 2014; 15: 16 – 18

Pulmonology
A Functional Anatomic Defect of the Cystic Fibrosis Airway
American Journal of Respiratory and Critical Care Medicine 2014; 190: 421 - 432

Bright-Thomas RJ., Johnson SC.
What is the role of noninvasive ventilation in cystic fibrosis?
Current Opinion In Pulmonary Medicine 2014; 20: 618 - 622

An observational study of matrix metalloproteinase (MMP)-9 in cystic fibrosis

Fischer AJ., Singh SB., Adam RJ., Stoltz DA., Baranano CF., Kao S., Weinberger MM., McCray PB., Stainer TD.
Tracheomalacia Is Associated With Lower FEV1 and Pseudomonas Acquisition in Children With CF
Pediatric Pulmonology 2014; 49: 960 - 970
Giron-Moreno RM., Justicia JL., Yamamoto S., Valenzuela C., Cisneros C., Gomez-Punter RM., Fernandes-Vasconcelos G., Ancochea J. Role of C-reactive protein as a biomarker for prediction of the severity of pulmonary exacerbations in patients with cystic fibrosis BMC Pulmonary Medicine 2014; 14: Arlo: 150


Hayes D., Tobias JD., Mansour HM., Kirkby S., McCoy KS., Daniels CJ., Whitlon BA. Pulmonary Hypertension in Cystic Fibrosis with Advanced Lung Disease American Journal Of Respiratory And Critical Care Medicine 2014; 190: 898 - 905

Illing EA., Woodworth BA. Management of the upper airway in cystic fibrosis Current Opinion In Pulmonary Medicine 2014; 20: 623 - 631


Nair C., Shoemark A., Chan M., Ollosson S., Dixon M., Hogg C., Alton EFWF., Davies JC., Williams HD. Cyanide levels found in infected cystic fibrosis sputum inhibit airway ciliary function European Respiratory Journal 2014; 44: 1253 - 1261


Savastano V., Bertin S., Vittori T., Tripodi C., Magliulo G. Evaluation of chronic rhinosinusitis management using the SNOT-22 in adult cystic fibrosis patients European Review for Medical and Pharmacological Sciences 2014; 18: 1985 - 1989


Taylor-Robinson DC., Thielen K., Pressler T., Olesen HV., Diderichsen F., Diggle PJ., Smyth R., Whitehead M. Low socioeconomic status is associated with worse lung function in the Danish cystic fibrosis population European Respiratory Journal 2014; 44: 1363 - 1366

Thamboo A., Santos RCD., Naidoo L., Rahmanian R., Chilvers MA., Chadha NK. Use of the SNOT-22 and UPSIT to Appropriately Select Pediatric Patients With Cystic Fibrosis Who Should Be Referred to an Otolaryngologist Cross-sectional Study JAMA Otolaryngology-Head & Neck Surgery 2014; 140: 934 - 939


Radiology

Bortoluzzi CF., Volpi S., D’Orazio C., Tiddens HAWM., Loeve M., Tridelio G., Assael BM. Bronchiectases at early chest computed tomography in children with cystic fibrosis are associated with increased risk of subsequent pulmonary exacerbations and chronic pseudomonas infection Journal of Cystic Fibrosis 2014; 13: 564 - 571


Pulmonary Disease in Cystic Fibrosis: Assessment with Chest CT at Chest Radiography Dose Levels
Radiology 2014; 272: 597 - 605

Hayes D., Long FR., McCoy KS., Sheikh SI.

Improvement in Bronchiectasis on CT Imaging in a Pediatric Patient with Cystic Fibrosis on Ivacaftor Therapy
Respiration 2014; 88: 345

Hayes D., Long FR., Ryan-Wenger NA., Sheikh SI.

Pulmonologist Versus Radiologist Interpretation of Cystic Fibrosis on CT Imaging
Lung 2014; 192: 637 - 638

Kuo WY., Ciet P., Tiddens H., Zhang W., Guillerman RP., van Straten M.

Cumulative Radiation Exposure to Abdominal Organs in Patients with Cystic Fibrosis Should Not Be Forgotten
American Journal Of Respiratory And Critical Care Medicine 2014; 190: 962

Locke LW., Myerburg MM., Markovetz MR., Parker RS., Weber L., Czechowski MR., Harding TJ., Brown SL., Nero JA., Pilewski JM., Corcoran TE.

Quantitative imaging of airway liquid absorption in cystic fibrosis
European Respiratory Journal 2014; 44: 675 - 684

Murphy KP., O’Connell OJ., O’Connor OJ., Plant BJ.

Cumulative Radiation Exposure to Abdominal Organs in Patients with Cystic Fibrosis Should Not Be Forgotten
American Journal of Respiratory and Critical Care Medicine 2014; 190: 961 - 962


Assessment of Repeatability of Hyperpolarized Gas MR Ventilation Functional Imaging in Cystic Fibrosis
Academic Radiology 2014; 21: 1524 - 1529

Tepper LA., Caudri D., Utens EMJW., van der Wiel EC., Quittner LA., Tiddens HAWM.

Tracking CF Disease Progression with CT and Respiratory Symptoms in a Cohort of Children Aged 6-19 Years
Pediatric Pulmonology 2014; 49: 1182 - 1189

Tiddens HAWM., Rosenow T.

What did we learn from two decades of chest computed tomography in cystic fibrosis?
Pediatric Radiology 2014; 44: 1490 - 1495

Ziai S., Coriati A., Chabot K., Mailhot M., Richter MV., Rabasa-Lhoret R.

Agreement of bioelectric impedance analysis and dual-energy X-ray absorptiometry for body composition evaluation in adults with cystic fibrosis

Screening


Public views on participating in newborn screening using genome sequencing
European Journal of Human Genetics 2014; 22: 1248 - 1254

Christiansen AL., Nybo M.

Lack of harmonization in sweat testing for cystic fibrosis - a national survey

Cortes E., Roldan AM., Palazon-Brus A., Rizo-Baeza MM., Manero H., Gil-Guillen VF.

Differences in immunoreactive trypsin values between type of feeding and ethnicity in neonatal cystic fibrosis screening: a cross-sectional study
Orphanet Journal of Rare Diseases 2014; 9: ArNo: 166


Human Genetics Society of Australasia Position Statement: Population-Based Carrier Screening for Cystic Fibrosis
Twin Research and Human Genetics 2014; 17: 578 - 583

Finan C., Nasr SZ., Rothwell E., Tarini BA.

Primary Care Providers’ Experiences Notifying Parents of Cystic Fibrosis Newborn Screening Results
Clinical Pediatrics 2015; 54: 67 - 75

Gusky S.

Students as Technicians: Screening Newborns for Cystic Fibrosis
American Biology Teacher 2014; 76: 254 - 258

Heidendaal JF., Tabbers MM., De Vreeze I.

False negative newborn screen and neonatal cholestasis in a premature child with cystic fibrosis

Massie J., Ioannou L., Delatycki M.

Prenatal and preconception population carrier screening for cystic fibrosis in Australia: Where are we up to?
Australian & New Zealand Journal of Obstetrics & Gynaecology 2014; 54: 593 - 599

Nguyen TTD., Thia LP., Hoo AF., Bush A., Aurora P., Wade A., Chudleigh J., Lum S., Stocks J.

Evolution of lung function during the first year of life in newborn screened cystic fibrosis infants
Thorax 2014; 69: 910 - 917


Benefits and burdens of newborn screening: public understanding and decision-making
Personalized Medicine 2014; 11: 593 - 607

Sengar AS., Agarwal A., Singh MK.

Cystic Fibrosis: Need for Mass Deployable Screening Methods
Applied Biochemistry and Biotechnology 2014; 174: 1127 - 1136

Serrano IC., Stoica G., Adams AM., Palomares E.

Dual core quantum dots for highly quantitative ratiometric detection of trypsin activity in cystic fibrosis patients
Nanoscale 2014; 6: 13623 - 13629

Therapy

Bilton D., Stanford G.

The expanding armamentarium of drugs to aid sputum clearance: how should they be used to optimize care?
Current Opinion In Pulmonary Medicine 2014; 20: 601 - 606
Bradley JM., Koker P., Deng Q., Moroni-Zentgraf P., Ratjen F., Geller DE., Elborn JS.
Testing Two Different Doses of Tiotropium Respimat (R) in Cystic Fibrosis: Phase 2 Randomized Trial Results
PLOS One 2014; 9: 8:e106195

Colombo C.
Mutation-targeted personalised medicine for cystic fibrosis
Lancet Respiratory Medicine 2014; 2: 863 - 865

d’Angelo I., Conte C., La Rotonda MI., Miro A., Quaglia F., Ungaro F.
Improving the efficacy of inhaled drugs in cystic fibrosis: Challenges and emerging drug delivery strategies
Advanced Drug Delivery Reviews 2014; 75: 92 - 111

De Boeck K., Munck A., Walker S., Faro A., Hiatt P., Gilmartin G., Higgins M.
Efficacy and safety of ivacaftor in patients with cystic fibrosis and a non-G551D gating mutation

Hadida S., Van Goor F., Dinehart K., Looker AR., Mueller P., Grootenhuis PDJ.
Case History: Kalydeco (R) (VX-770, Ivacaftor), a CFTR Potentiator for the Treatment of Patients with Cystic Fibrosis and the G551D-CFTR Mutation
Annual Reports in Medicinal Chemistry 2014; 49: 383 - 398

Hayes D., McCoy KS., Sheikh SI.
Improvement of Sinus Disease in Cystic Fibrosis with Ivacaftor Therapy
American Journal of Respiratory and Critical Care Medicine 2014; 190: 468

Hurt K., Bilton D.
Inhaled Interventions in Cystic Fibrosis: Mucoactive and Antibiotic Therapies
Respiration 2014; 88: 441 - 448

Kopeikin Z., Yuksek Z., Yang HY., Bompadre SG.
Combined effects of VX-770 and VX-809 on several functional abnormalities of F508del-CFTR channels
Journal of Cystic Fibrosis 2014; 13: 508 - 514

Kumar S., Tana A., Shankar A.
Cystic fibrosis - What are the prospects for a cure?
European Journal of Internal Medicine 2014; 25: 803 - 807

Lamas A., Maiz L., de Valbuena MR., Gonzalez-Casbas JM., Suarez L.
Subcutaneous implant with etonogestrel (Implanon (R)) for catamenial exacerbations in a patient with cystic fibrosis: a case report
BMC Pulmonary Medicine 2014; 14: ArNo: 165

Liu ZC., Borlak J., Tong WD.
Deciphering miRNA transcription factor feed-forward loops to identify drug repurposing candidates for cystic fibrosis
Genome Medicine 2014; 6: ArNo: 94

Mainz JG., Michel R., Arnold C.
Nasal saline as a placebo in chronic rhinosinusitis Response
Journal of Cystic Fibrosis 2014; 13: 602 - 603

Markovetz MR., Corcoran TE., Locke LW., Myerburg MM., Pilewski JM., Parker RS.
A Physiologically-Motivated Compartment-Based Model of the Effect of Inhaled Hypertonic Saline on Mucociliary Clearance and Liquid Transport in Cystic Fibrosis

McColley SA.
Ivacaftor therapy for cystic fibrosis
Expert Opinion on Orphan Drugs 2014; 2: 1225 - 1232

McKone EF., Borowitz D., Drevinek P., Griese M., Konstan MW., Wainwright C., Ratjen F., Sermet-Gaudelus I., Plant B., Munck A., Jiang Y., Gilmartin G., Davies JC.
Long-term safety and efficacy of ivacaftor in patients with cystic fibrosis who have the Gly551Asp-CFTR mutation: a phase 3, open-label extension study (PERSIST)
Lancet Respiratory Medicine 2014; 2: 902 - 910

O’Neill DA., Fraser-Pitt D.
Progress towards next-generation therapeutics for cystic fibrosis
Future Medical Chemistry 2014; 6: 1067 - 1079

Peled O., Kalamarov V., Kerem E., Shoseyov D., Blau H., Efrati O., Block C.
Contamination of hypertonic saline solutions in use by cystic fibrosis patients in Israel
Journal of Cystic Fibrosis 2014; 13: 550 - 556

Reznikov LR., Abou Aliawa MH., Doehrn CL., Gansemere ND., Diekema DJ., Stoltz DA., Welsh MJ.
Antibacterial properties of the CFTR potentiator ivacaftor
Journal of Cystic Fibrosis 2014; 13: 515 - 519

Robertson SM., Luo X., Dubey N., Li CH., Chavan AB., Gilmartin GS., Higgins M., Mahnke L.
Clinical Drug-Drug Interaction Assessment of Ivacaftor as a Potential Inhibitor of Cytochrome P450 and P-glycoprotein

Roomans GM.
Pharmacological treatment of the basic defect in cystic fibrosis
Cell Biology International 2014; 38: 1244 - 1246

Saynor ZL., Barker AR., Oades PJ., Williams CA.
The Effect of Ivacaftor in Adolescents With Cystic Fibrosis (G551D Mutation): An Exercise Physiology Perspective
Pediatric Physical Therapy 2014; 26: 454 - 461

Shah N., Perrin F.
Extracorporeal membrane oxygenation (ECMO) in cystic fibrosis
Paediatric Respiratory Reviews 2014; 15: 26 - 28

Pooled analysis of tiotropium Respimat (R) pharmacokinetics in cystic fibrosis who have the Gly551Asp CFTR mutation

Syed BA., Hamad B.
The cystic fibrosis drug market

Tait BD., Miller JP.
Disease-Modifying Agents for the Treatment of Cystic Fibrosis
Annual Reports In Medicinal Chemistry 2014; 49: 317 - 330

Toltzis P.
A 5-Year Clinical Evaluation of Therapeutic Program for Patients with Cystic Fibrosis
Journal of Pediatrics 2014; 165: 966

Wainwright CE.
Ivacaftor for patients with cystic fibrosis
Expert Review of Respiratory Medicine 2014; 8: 533 – 538

Urology

Korzeniowska-Eksterowicz A., Stelmach J., Stelmach W.
Urinary incontinence in adolescent females with cystic fibrosis in Poland
Central European Journal of Medicine 2014; 9: 778 - 783

Sener D., Cokugras H., Camcioglu Y., Aakkaya N., Kilicaslan I., Sevler L., Canpolat N.
Renal Amyloidosis Secondary to Cystic Fibrosis
Indian Journal of Pediatrics 2014; 81: 973 - 974