

2020 ECFS Conference

New Frontiers in Basic Science of Cystic Fibrosis

25-28 March 2020, Albufeira, Portugal

Programme

Chairpersons:

Carlos Farinha (Lisbon, PT), Nicoletta Pedemonte (Genoa, IT) Jeffrey L. Brodsky
(Pittsburgh, US)

Wednesday, 25 March 2020 (Day 1)

- 13:30-17:00** **Pre-Conference Seminar (organised by the ECFS and the Patient organisations)**
Antimicrobial resistance in cystic fibrosis
Chairs: Jane Davies (UK) Pavel Drevinek (CZ)
[Programme](#)
- 17:30-18:00** **Official Opening of the Meeting by the Conference Chairpersons**
- 18:00-19:00** **Opening Keynote Lecture**
Studies in the pig airway to model CF - Lynda S. Ostedgaard (US)
- 19:00-19:45** **Welcome Reception**
- 19:45-21:30** *Dinner*

Thursday, 26 March 2020 (Day 2)

07:30-08:45 *Breakfast*

08:45-10:30 Symposium 1 – From genotype to phenotype

Chairs: Martina Gentzsch (US) - Isabelle Sermet-Gaudelus (FR)

- 08:45-09:10 Complex genetics and penetrance of variants in a monogenic disorders: cystic fibrosis as a model disorder - Milan Macek (CZ)
- 09:10-09:35 Perspectives on precision therapeutics for cystic fibrosis lung disease - Eric Sorscher (US)
- 09:35-10:00 Rare mutations in cystic fibrosis: from molecular diagnosis to clinical applications - Felice Amato (IT)
- 10:00-10:10 Abstract 13 - Analysis of aberrant splicing patterns of CFTR variants with premature termination codons and other disease-causing mutations - Normand Allaire (US)
- 10:10-10:20 Abstract 16 - LncRNAs: emerging players in CFTR gene regulation - Jessica Varilh (FR)
- 10:20-10:30 Abstract 03 - Using targeted locus amplification to study heterogeneity within the CFTR F508del gene – Juliet Lefferts (NL)

10:30-11:00 *Coffee break & Poster viewing*

11:00-12:45 Symposium 2 – CFTR (i): from structure to function

Chairs: Christine Bear (CA) - Carlos Farinha (PT)

- 11:00-11:25 Specific evolution of the CFTR sequence in the ABC superfamily: implication for modulation- Isabelle Callebaut (FR)
- 11:25-11:50 CFTR and ABC transporter structures: drug binding sites - Bob Ford (UK)
- 11:50-12:15 Conditional non-hydrolytic gating by hydrolysable ATP - T.C. Hwang (US)
- 12:15-12:25 Abstract 21 - Can two wrongs make a right? Studies on F508-del-CFTR revertant mutations – Stella Prins (UK)
- 12:25-12:35 Abstract 26 - CFTR-France: a comprehensive web resource for exploring CF genetics and CFTR 3D structure - Souphatta Sasorith (FR)
- 12:35-12:45 Abstract 17 - Structural effects of a pathogenic CFTR loop mutation and its dynamic stabilization by Lumacaftor - Mathias Schenkel (DE)

12:45-14:30 *Lunch*

14:30-15:30 Flash Poster Session (even numbers)

Chair: Bob Ford (UK)

15:30-16:00 *Coffee break & Poster viewing*

16:00-17:45 Symposium 3 – CFTR (ii): Trafficking and Interactions

Chairs: Jeff Brodsky (US) - Agnes Swiatecka-Urban (US)

- 16:00-16:25 Requirements for CFTR folding and transport - Ineke Braakman (NL)
- 16:25-16:50 CFTR interactome: the role of plasma membrane lipids - Massimo Aureli (IT)
- 16:50-17:15 Pancreas on a chip: Personalized model system for individual CF-patients - AP Naren (US)
- 17:15-17:25 Abstract 45 - iPSC derived intestine organoids model defective F508del-CFTR function in vitro - Sunny Xia (CA)

17:25-17:35 Abstract 27 - Elucidating the mechanism for CFTR plasma membrane stabilization under EPAC1 activation - João F. Ferreira (PT)

17:35-17:45 Abstract 37 - Proteomic profiling of F508del-CFTR cells to identify new pharmacological targets for CF – Clarissa Braccia (IT)

17:45-18:00 *Break*

18:00-19:45 **ECFS Basic Science Working Group session**

Personalized Medicine Approaches to Treat CF

19:45-21:30 *Dinner*

21:30-23:00 **Evening Poster Session:** Posters with Even numbers

Friday, 27 March 2020 (Day 3)

07:30-08:45 *Breakfast*

08:45-10:30 Symposium 4 – Other channels that modify CFTR containing epithelia

Chairs: TC Hwang (US) - Nicoletta Pedemonte (IT)

- 08:45-09:10 Calcium channels, inflammation and CF- Rob Tarran (US)
- 09:10-09:35 Expression and function of ATP12A protein in airway epithelial cells - Luis Galiotta (IT)
- 09:35-10:00 TMEM16A potentiation, a novel approach for the treatment of CF - Martin Gosling (UK)
- 10:00-10:10 Abstract 50 - Targeting alternative chloride channels in CF - Madalena C. Pinto (PT)
- 10:10-10:20 Abstract 49 - Furin inhibition as a mechanism to reduce aberrant ENaC-mediated sodium transport and rehydrate the airways in cystic fibrosis lung disease - Lisa E. J. Douglas (UK)
- 10:20-10:30 Abstract 54 - Transmembrane water fluxes regulated by CFTR in living human airway epithelial CFBE cells and CHO cells. Effect of VX770 and VX809 on G551D and F508del-CFTR – Frederic Becq (FR)

10:30-11:00 *Coffee break & Poster viewing*

11:00-12:45 Symposium 5 - CFTR (iii): Beyond the airway

Chairs: Mike Gray (UK) - Ineke Braakman (NL)

- 11:00-11:25 CFTR: a new horizon in pancreatitis - Peter Hegyi (HU)
- 11:25-11:50 Strategies to improve gut fluidity and alkalinity in the CF gut beyond CFTR rescue - Ursula Seidler (DE)
- 11:50-12:15 Defective renal bicarbonate excretion in cystic fibrosis - Karl Kunzelmann (DE)
- 12:15-12:25 Abstract 63 - Linking CFTR to epithelial differentiation by functional genomics - Margarida C. Quaresma (PT)
- 12:25-12:35 Abstract 65 - Evaluation of ionocytes abundance and CFTR expression in human airways epithelia - Paolo Scudieri (IT)
- 12:35-12:45 Abstract 55 - The Wnt signaling pathway is deregulated in cystic fibrosis airway epithelium - Tahir Idris (CH)

12:45-14:00 *Lunch*

14:00-18:30 **Free Afternoon**

18:30-19:30 Flash Poster Session (odd numbers)

Chair: Massimo Aureli (IT)

19:30 -21:30 *Dinner*

21:30-23:00 Evening Poster Session: Posters with Odd numbers

Saturday, 28 March 2020 (Day 4)

07:30-08:45 *Breakfast*

08:45-10:30 **Symposium 6 – Inflammation and infection**

Chairs: Rob Tarran (US) - Martin Mense (US)

- 08:45-09:10 A fishy solution: understanding the immune pathogenesis in CF through zebrafish models - Audrey Bernut (UK)
- 09:10-09:35 Novel mechanisms of TGF-beta signaling in the airway and relevance to cystic fibrosis - Agnes Swiatecka-Urban (US)
- 09:35-10:00 3 to 5 years evolution of the lung microbiome in adolescents with cystic fibrosis - Sébastien Boutin (DE)
- 10:00-10:10 Abstract 69 - Bridging CFTR-modulators to Pseudomonas aeruginosa infection and antibiotic efficacy - Cristina Cigana (IT)
- 10:10-10:20 Abstract 70 - Functional and phenotypic alterations of phagocytes in newborn CFTR-/- pigs - Florian Jaudas (DE)
- 10:20-10:30 Abstract 67 - Transcriptomic analysis of CFTR-deficient endothelial cells reveals a pro-inflammatory phenotype - Mathias Declercq (BE)

10:30-11:00 *Coffee break & Poster viewing*

11:00-12:45 **Symposium 7 – Mucus and mucins**

Chairs: Karl Kunzelmann (DE) – Ursula Seidler (DE)

- 11:00-11:25 CF mucus: how to reverse aberrant properties - Camille Ehre (US)
- 11:25-11:50 What is wrong with the CF lung mucus and how can we fix it? - Gunnar Hansson (SE)
- 11:50-12:15 Building and testing a live model of mucociliary clearance using the Xenopus tropicalis tadpole - Eamon Dubaissi (UK)
- 12:15-12:25 Abstract 94 - Intramolecular interactions of the airway mucin, MUC5B and their role in packaging within secretory granules - Caroline Ridley (UK)
- 12:25-12:35 Abstract 97 - Airway macrophages in a mouse model of mucociliary clearance deficiency show a dysregulated epigenome - Michelle Paulsen (DE)
- 12:35-12:45 Abstract 100 - Evaluation of the physiological role of the TMEM16A chloride channel in airway epithelia - Arianna Venturini (IT)

12:45-14:15 *Lunch*

14:15-16:00 Symposium 8 – Current challenges in therapeutics

Chairs: Kris De Boeck (BE) - Eric Sorscher(US)

14:15-14:40 What is missing? Lessons from patient care - Isabelle Sermet-Gaudelus (FR)

14:40-15:05 Dissecting the potency of mesoangioblasts for an innovative cell-based therapy for cystic fibrosis disease - Graziella Messina (IT)

15:05-15:30 Evaluation of efficacy of pharmacological rescue of F508del and other misfolding mutations - Martina Gentzsch (US)

15:30-15:40 Abstract 117 - Delivery of CRISPR/Cas9 by receptor-targeted nanocomplexes as a potential therapy for cystic fibrosis - Amy Walker (UK)

15:40-15:50 Abstract 119 - Pharmacological rescue of missense and nonsense mutation function with triple CFTR modulators - Ashvani Singh (US)

15:50-16:00 Abstract 120 - Novel amplifier-enhanced high-throughput screen targeting CFTR nonsense mutation suppression identifies translational read-through modulators with different profiles - Danijela Dukovski (US)

16:00-16:30 *Coffee Break*

16:30-17:30 Workshop: Tools for Research

Chairs: Luis Galletta (IT)

16:30 – 16:50 ABCM2: the web application for predicting the effect of mutations in ABC proteins - Tamas Hegedus (HU)

16:50 – 17:10 Looking at molecular landscapes for multi-target profiling of new prospective drugs - André Falcão (PT)

17:10 – 17:30 CFFT Lab: tools available to the research community – Martin Mense (US)

18:00-19:00 Closing Keynote lecture

Striking CF by mechanistic approaches – Margarida Amaral (PT)

20:30 *Dinner / Social Event*