



Fondazione per la Ricerca
sulla Fibrosi Cistica - Onlus
italian cystic fibrosis research foundation

**XIX Convention of Investigators in cystic fibrosis
(webinar)
25-26 November, 2021
Camera di Commercio di Verona, congress center**

Final Program

Thursday, November 25th

10.00 *Opening*

10.05 *Memento of Prof. Gianni Mastella*

10.15 *Greetings*

- Matteo Marzotto (*President FFC Ricerca*)
- Gianna Puppo Fornaro (*President LIFC Lega Italiana Fibrosi Cistica*)
- Marco Cipolli (*President SIFC Società Italiana Fibrosi Cistica*)
- Callisto Marco Bravi (*Director AOUI Verona*)
- Giorgio Berton (*Scientific director FFC Ricerca*)

10.35 – 12.10

Session 1

MODULATION OF CFTR FUNCTION

Chair Armirotti A, Volpi S

1. Galietta L

Identification of deubiquitinases and ubiquitin ligases that affect mutant CFTR rescue (FFC#6/2019, concluded) (15')

2. Netti P, Di Bernardo D

Investigating epithelial-stromal crosstalk in full thickness cystic fibrosis model on chip for evaluating novel therapeutic strategies (FFC#14/2019, concluded) (15')

3. Salvi M

Functional role of post-translational modifications in F508del-CFTR correction (FFC#7/2020, concluded) (15')

4. Bertozzi F, Romeo E

Characterization of CFTR modulators mechanism of action via Photo-Affinity Labeling (PAL) approach (FFC#4/2020, ongoing) (15')

5. Bragonzi A, Rossi G

Bridging airway mucus-microbiota-host genotype to define novel cystic fibrosis models (FFC#2/2019, concluded) (15')

11.50 – 12.10 *Discussion*

12.10 – 13.30

Session 2

**RECOVERY OF CFTR FUNCTION BY POTENTIATORS/CORRECTORS:
EX VIVO STUDIES**

Chair: Barraja P, Volpi S

6. Pedemonte N, Cavalli A

Therotyping orphan mutations in Italian cystic fibrosis patients: efficacy of CFTR modulators and RNF5 inhibitors (FFC#9/2019, concluded) (15')

7. Eramo A, Lucarelli M

Establishment of conditionally reprogrammed airway epithelial stem cell cultures from nasal epithelia of cystic fibrosis patients: exploring response to CFTR-modulating drugs for correlation with genetic profile (therotyping) and restoring CFTR function through gene editing approaches (FFC#8/2020, concluded) (15')

8. Frulloni L, de Jonge H, Lucidi V

Intestinal organoids for assessment and pharmacological correction of abnormalities in fluid transport and anion currents in patients affected by pancreatitis (FFC#5/2020, concluded) (15')

9. Melotti P, de Jonge H, Castaldo G

Therotyping of rare CFTR genotypes for treatment with CFTR modulators (FFC#9/2020, ongoing) (15')

13.10 – 13.30 *Discussion*

13.30 – 14.30 *Lunch (brown-bag)*

14.30 – 16.55

Session 3

NEW STRATEGIES TO RECOVER MUTATED CFTR FUNCTION

Chair: Colombo C, Bandiera T

10. Barraja P, Venturini A

Small nitrogen heterocycles as correctors of the mutant CFTR protein in cystic fibrosis (FFC#3/2020, ongoing) (15')

11. Cereseto A, Arosio D

Harnessing CRISPR/Cas technology to revert DF508 CFTR defect (FFC#3/2019, concluded + extension 2021) (15')

12. Lentini L, Pibiri I

Validation of the distribution and activity of new optimized leads in mouse model and other CF model systems (FFC#6/2020, ongoing) (15')

15.15 – 15.35 *Discussion*

13. Amato F

Peptide-nucleic acids as potential CFTR amplifier molecules for cystic fibrosis treatment (FFC#1/2020, ongoing) (15')

14. Mangoni M

Antimicrobial peptides from amphibian skin for treatment of lung pathology in cystic fibrosis: advanced *in vitro* and *in vivo* functional characterization (FFC#8/2019, concluded) (15')

15. Aureli M, Tamanini A

Lipid-based therapeutic strategies to optimize the effectiveness of innovative drugs to rescue F508del-CFTR (FFC#2/2020, ongoing) (15')

16. Cozza G, Esposito S, Raia V

Restoring defective proteostasis in cystic fibrosis: novel strategies for F508del-CFTR repair (FFC#4/2019, concluded) (15')

16.35 – 16.55 *Discussion*

16.55 – 17.15 *Coffee break*

17.15 – 17.55

Carla Colombo – *Special Lecture*

From the bench to the bedside: focus on the effects of the new modulator/corrector therapies in patients' real life (40')

17.55 – 18.05 *Discussion moderated by Carlo Castellani*

19.45 - ***Welcome dinner***

Friday, November 26th

8.30 – 11.25

Session 4

**INFECTIONS: EMERGING PATHOGENS AND
NEW THERAPEUTIC STRATEGIES**

Chair Bragonzi A, Mencarini J

Nontuberculous Mycobacteria (NTM) Infections

17. Fraziano M

Preclinical study of a combined host- and pathogen directed approach based on bioactive liposomes and bacteriophages against *Mycobacterium abscessus* infection (FFC#21/2019, concluded) (15')

18. Lorè NI

Unravelling novel biomarkers to define the progression of *Mycobacterium abscessus* lung disease in cystic fibrosis (FFC#23/2020, ongoing) (15')

19. Pasca MR, Makarov V, Ramón-García S, Tortoli E

New weapons against *Mycobacterium abscessus* and other nontuberculous mycobacteria (FFC#14/2020, pilot project concluded, FFC#18/2021 extension) (15')

20. Fattorini L, Borroni E

New drug combinations against non-tuberculous mycobacteria infections in cystic fibrosis (FFC#12/2020, pilot project concluded, FFC#17/2021, extension) (15')

9.30 – 9.50 *Discussion*

9.50 – 11.25

New antimicrobial strategies

21. Lleo' M

Investigating *Achromobacter xylosoxidans* pathogenicity and clinical role in CF lung infection (FFC#18/2019, concluded) (15')

22. Visca P, Sorrentino R

Gallium as an antibacterial agent in cystic fibrosis: animal studies for the delivery of inhalable formulations to the clinic (FFC#19/2019, concluded) (15')

23. Cellini B

Exploring the dual targeting of host and microbial sphingosine-1-phosphate lyase as antimicrobial strategy in cystic fibrosis (FFC#16/2020, concluded, FFC#19/2021, extension) (15')

24. Guaragna A, De Gregorio E

Can old and new sweet glycomimetics act as antibacterial and antibiofilm agents in the treatment of CF lung disease infections? (FFC#13/2020, ongoing) (15')

25. Piacentini M, Raia V

Targeting the STING/Transglutaminase 2-regulated Interferon response as a novel host-direct approach to fight bacterial infections in cystic fibrosis (FFC#15/2020, ongoing) (15')

11.05 – 11.25 *Discussion*

11.25 – 11.45 *Coffee break*

11.45 – 13.35

Session 5

CLINICAL RESEARCH

Chair: Cipolli M, Braggion C

26. Battezzati A, Colombo C, Lucidi V, Lucanto MC, Mari A

Early derangements of glucose tolerance in cystic fibrosis: effect of CFTR modulators (FFC#24/2019, concluded) (15')

27. Morana G, Colzani G

Standardized Ventilation Inflammation Perfusion and Structure (VIPS) MRI platform for monitoring cystic fibrosis lung disease (FFC#26/2019, concluded) (15')

28. Scaravilli V

Right ventricle dysfunction in cystic fibrosis patients undergoing lung transplant (FFC#27/2019, ongoing) (15')

29. Aliverti A

Use of multivolume MRI to assess response to CFTR modulators (FFC#21/2020, concluded) (15')

30. Cirilli N, Tiano L, Gesuita R

Role of viable but non culturable (VBNC) bacterial forms in CF patients in a clinical setting: a translational research (FFC#22/2020, ongoing) (15')

31. Terlizzi V, Tosco A, Claut LE

Cystic fibrosis screen positive inconclusive diagnosis (CFSPID): an Italian multicenter survey evaluating prevalence, clinical data, management and outcome (FFC#24/2020, ongoing) (15')

13.15 – 13.35 *Discussion*

13.35 – 14.35 *Lunch (brown-bag)*

14.35 – 15.55

Session 6

INFECTIONS: *P. AERUGINOSA*

Chair: Pasca MR, Delfino E

32. Ascenzioni F, Imperi F, Botta B

Pharmacological inhibition of colistin resistance in gram-negative cystic fibrosis pathogens (FFC#15/2019, concluded) (15'), *online presentation*

33. Biavasco F, Citterio B, Mangiaterra G

Fighting *Pseudomonas aeruginosa* persists in cystic fibrosis pulmonary infections: improved detection and therapeutic strategies (FFC#16/2019, concluded) (15')

34. Bertoni G

Targeting small RNA-mediated regulation of virulence and antibiotic resistance to develop non-traditional therapeutic options against *Pseudomonas aeruginosa* (FFC#10/2020, ongoing) (15')

35. Brun P, Marzaro G

Disrupting *Pseudomonas aeruginosa* Quorum Sensing signalling in cystic fibrosis patients as a new frontier for antibacterial therapy (FFC#11/2020, concluded) (15')

15.35 – 15.55 *Discussion*

15.55 – 17.15

Session 7

INFLAMMATION

Chair: Cabrini G, Minicucci L

36. Recchiuti A, Aloisi A

Nanotechnology-based Resolvin D1 as proresolving therapy in cystic fibrosis: preclinical studies for the delivery of innovative formulations to the clinic (FFC#19/2020, pilot project concluded, FFC#20/2021 extension)

37. Giovagnoli S

Oral and pulmonary delivery platforms for anakinra repurposing in cystic fibrosis (FFC#17/2020, ongoing)

38. Summa V, Altucci L

Harnessing selective histone deacetylase 6 (HDAC6) inhibition to tackle inflammation and fibrotic remodeling in cystic fibrosis (FFC#20/2020, concluded)

39. Paroni M, Johansen HK

Counteracting inflammation triggered by *P. aeruginosa*-activated lung-infiltrating Th1/17 cells: a novel approach for precision medicine in CF. (FFC#18/2020, ongoing)

16.55 – 17.15 *Discussion*

17.15 – 17.20 *Closing remarks*