

Milan
5-6 September
2019

Università degli Studi di Milano



1st Adult Cystic Fibrosis International Workshop



Adult Cystic Fibrosis

FINAL PROGRAMME



12

EBAP
credits
GRANTED

13

EUROPEAN CME
credits (ECMEC®S)
GRANTED


AFISM
Association Internationale pour la Promotion de Formations
Spécialisées en Médecine et en Sciences Biologiques

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www.adultcysticfibrosis.org



President

Francesco Blasi (Italy)

Scientific and Organising Committee

Stefano Aliberti (Italy)

Carla Colombo (Italy)

Stuart Elborn (UK)

Paolo Palange (Italy)

Valeria Raia (Italy)

Michal Shteinberg (Israel)



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welcome letter

Dear Colleagues,

Welcome to the **1st Adult Cystic Fibrosis International Workshop**.

In the past thinking of dedicating a whole International Congress to Adult Cystic Fibrosis would have been considered as an utopia, but, in the last decades, the new epidemiology and the advancement in diagnosis, therapies and treatments have made this topic worth of a dedicate and deep attention.

The aim of the project is to gather the researchers and clinicians specialized in cystic fibrosis to discuss in depth the critical issues connected with the management of the pathology in adult age.

Thanks to the important goals reached in the last years and looking at the new perspectives for life management of patients with Cystic Fibrosis, we look forward that this meeting can become a traditional appointment, to support always new approaches, treatments and frontiers for Cystic Fibrosis.

Joining forces, exchanging clinical experiences, sharing data and instruments, creating a network of active experts in the field are the main goals of the conference.

For this reason, I wish you two great working days in Milan, where sharing knowledge and experiences will be the key values of our **1st Adult Cystic Fibrosis International Workshop**.

Yours Sincerely,

Francesco Blasi



Professor of Respiratory Medicine,
Department of Pathophysiology and Transplantation, Università degli Studi di Milano.
Member of Board of Directors, Università degli Studi di Milano.
Head Internal Medicine Department, Respiratory Unit and Adult Cystic Fibrosis Center.
Fondazione IRCCS Cà Granda Ospedale Maggiore Policlinico Milano.



president



Francesco Blasi

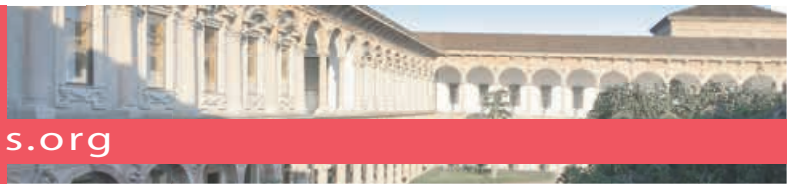
Francesco Blasi, MD, FERS is Professor of Respiratory Medicine in the Department of Pathophysiology and Transplantation at the University of Milan, Italy, as well as Head of Internal Medicine Department and Respiratory Unit of the Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy. He is also director of the Adult Cystic Fibrosis Center of the Lombardia Regional Referral CF Center and currently a member of the Board of Directors of the University of Milan.

From 2012–2013, he was president of European Respiratory Society (ERS) and from 2016–2017 president of the Italian Respiratory Society (SIP/IRS). From 2017–2018, he was president of the Italian Respiratory Society Research Center (SIP/IRS Centro Ricerche).

Professor Blasi has published more than 350 papers in international journals (January 2019: h-index: 67, Citations: 18,268).

His research interests include pneumonia, COPD, bronchiectasis, tuberculosis and NTM infections, cystic fibrosis and lung transplantation.

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Stefano Aliberti

Stefano Aliberti is Associate Professor in Respiratory Medicine at the University of Milan and Respiratory Consultant at the Policlinico University Hospital, Milan, Italy.

He received his medical degree from the University of Milan and completed his clinical research training at the University of Louisville, KY, USA, working as member of the Community-Acquired Pneumonia Organization Database.

Most of his researches over the past fifteen years have been focused on both acute and chronic respiratory infections, and particularly on community-acquired pneumonia, bronchiectasis and non-tuberculous mycobacteria (NTM).

He is former Head of the Respiratory Infections Assembly of the European Respiratory Society, chair of the European Registry of Bronchiectasis (EMBARC), the Italian Registry of Bronchiectasis (IRIDE) and the Italian Registry of pulmonary NTM (IRENE).



Carla Colombo

Carla Colombo is Professor of Pediatrics at the University of Milan and Director of the Regional Cystic Fibrosis Center, at Fondazione IRCCS Ca' Granda. Ospedale Maggiore Policlinico, Milan. She was promoting member and then President of the Italian Cystic Fibrosis Society (2007 – 2010).

Prof. Colombo has devoted most of her research activity to Cystic Fibrosis, including original studies on the natural history and treatment of CF-associated liver disease, CF-related diabetes, various nutritional issues also co-authoring the last European guidelines.

She has participated to the EuroCareCF project (European Coordination Action for Research in Cystic Fibrosis), as part of the Sixth Framework Program as Leader of Workpackage 1 (Optimizing Patient Care and Team Work), and later to the Horizon 2020 project MyCyFAPP (Innovative approach for self-management and social welfare of Cystic Fibrosis patients in Europe) which led to generation of a self-management app provided with a professional web tool for pancreatic enzyme replacement therapy. Since 2012, the CF Center of Milan is part of the Clinical Trial Network of the European Cystic Fibrosis Society (www.ecfs.eu.ctn), and Prof. Colombo has been PI and also national coordinator in many clinical trials carried out within this network.

She is author of more than 250 publications in international journals.



scientific and organising committee



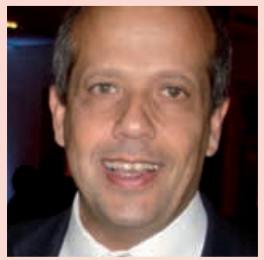
Stuart Elborn

Stuart Elborn, M.D., FRCP, is clinical professor of respiratory medicine, and centre director for specialist adult cystic fibrosis Royal Brompton Hospital in London.

Prof. Elborn has been Professor of Respiratory Medicine at Queens' University Hospital in Belfast and Dean of the School of Medicine, Dentistry and Biomedical Sciences at Queens University, Belfast until 2016. He has been Non-Executive Director at Belfast Health & Social Care Trust since April 1st, 2016.

Prof. Stuart Elborn trained in Belfast and helped developing Cystic Fibrosis Centres for adults in Nottingham and Cardiff. Back in Belfast in 1995 he started a new Adult Cystic Fibrosis Centre providing care for all adults with cystic fibrosis in Northern Ireland.

He was President of the European Cystic Fibrosis Society from 2008 to 2015.



Paolo Palange

Prof. Paolo Palange is Director of Pulmonary Function Unit (2001- today) and Director Division of Internal Medicine and Respiratory Medicine (2010-today) at Policlinico Umberto I – La Sapienza University of Rome.

He is Full Professor of Respiratory Medicine (2013-today) at La Sapienza University (Rome).

His main research fields are Cystic Fibrosis, COPD, Asthma, Lung Function Testing, Cardiopulmonary exercise testing, Exercise induced asthma.

He has been involved in the European Respiratory Society from the '90's. He has been chairmen of different ERS Task Forces, Seminar and Projects. He was the director of the ERS Hermes Program from 2011 to 2013.

He published more than 130 international articles and scientific works.

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Valeria Raia

Valerie Raia graduated in Medicine in 1979 and specialized in Paediatrics in 1983. Since 2011 she has been Associated Professor of Paediatrics at Federico II University in Naples, Italy.

She is Director of the Regional Center for Cystic Fibrosis, Pediatric Section in the Naples area. She started her research path on cystic fibrosis in 1986, when she started studying on CF pathogenesis and related issues.

She published more than 150 international articles and scientific works.

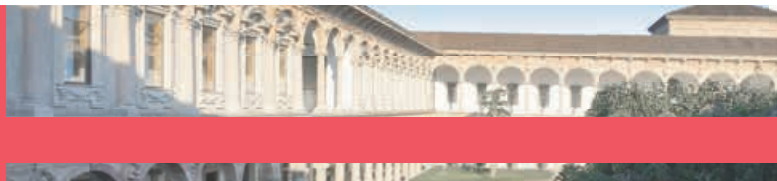
She has been elected President of the Italian Society for Cystic Fibrosis Study in 2017.



Michal Shteinberg

Michal Shteinberg is heading the bronchiectasis and adult CF service in the Pulmonology institute and CF center, Carmel medical center, Haifa, Israel.

She is a clinical lecturer in the Faculty of medicine at the Technion, Israel Institute of Technology. Her main research interests are bronchiectasis and its overlap with asthma and chronic rhinosinusitis, and adult CF- mainly, female infertility in CF.



faculty list

Name	Nationality
S. Aliberti	Italy
J. Altenburg	The Netherlands
B. Assael	Italy
C. Benden	Switzerland
F. Blasi	Italy
P. R. Burgel	France
C. Castellani	Italy
F. Cathcart	UK
S. Ceri	Italy
S.H. Chotirmall	Singapore
C. Colombo	Italy
M. Contarini	Italy
J. Davies	UK
K. de Winter - de Groot	The Netherlands
L. Dupont	Belgium
S. Elborn	UK
A. Gramegna	Italy
C. S. Haworth	UK
S. Madge	UK
J. Maetz	France
P. Maisonneuve	Italy
V. Makhmutova	Russian Federation
L. Morlacchi	Italy
P. Palange	Italy
B. Plant	Ireland
V. Raia	Italy
C. Schwarz	Germany
M. Shteinberg	Israel
N. J. Simmonds	UK
G. Taccetti	Italy
J. Wood	UK

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The **1st Adult Cystic Fibrosis International Workshop** is organized by:



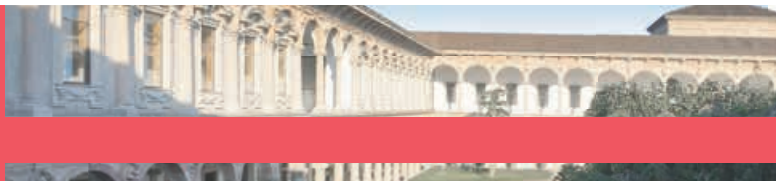
L'Association Internationale pour la promotion de Formations Spécialisées en Médecine et en Sciences Biologiques (AFISM) is an international association devoted to promoting and enhancing the continuing professional development and post-degree education in medical, scientific and technical professions. AFISM organises high level educational events, practical workshops, debates, consensus conferences and international congresses to help specialists to keep themselves up to date with all the new and quick advancements in the different disciplines.

Encouraging the networking among professionals, AFISM is also focused in strongly support the sharing of evidence-based knowledge and discourages the diffusion of practices which are not supported by evidence, preventing the circulation of harmful fake news but still maintaining an open dialogue with all the different actors.

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Adult Cystic Fibrosis



topics

- Cancer prevention
- CF Epidemiology
- Diagnosis overview
- End of life management
- ERS/ECFS/NICE guidelines in adult patients
- Lung transplant
- Management of infections
- New treatments for Cystic Fibrosis
- NTM: how to deal with
- Patients life management
- Pregnancy in Cystic Fibrosis patients
- Transition

venue



The **1st Adult Cystic Fibrosis International Workshop** is held in the prestigious University of Milan:

Università degli Studi di Milano

Via Festa del Perdono 7

Tel. +39 02 503.12103

For more information, please visit the website:

www.unimi.it

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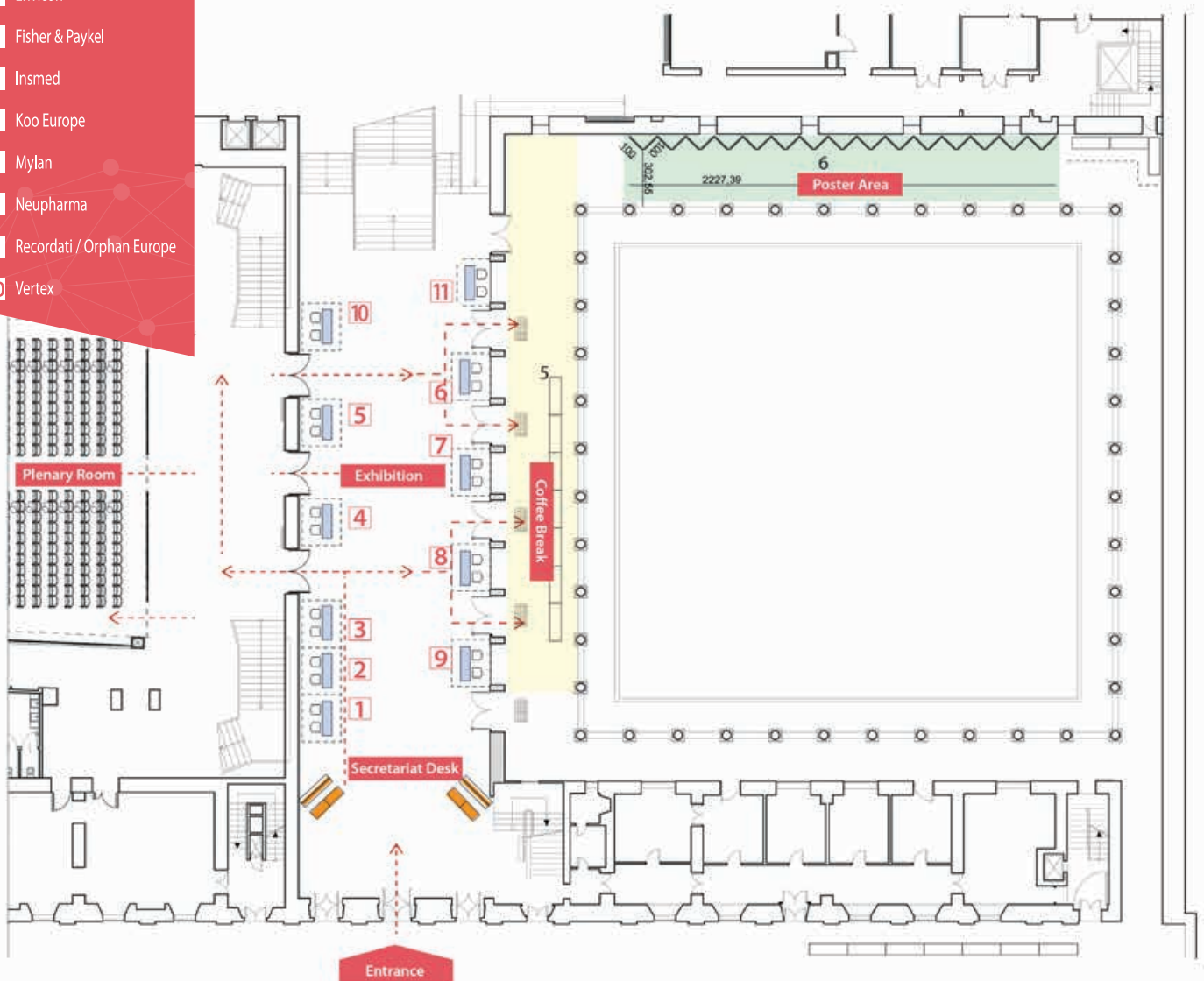




meeting spaces and exhibition layout

Booth Legend

- 1 Chiesi Group
- 2 Corbus Pharmaceuticals
- 11 Cosmed
- 4 Envicon
- 9 Fisher & Paykel
- 8 Insméd
- 5 Koo Europe
- 7 Mylan
- 3 Neupharma
- 6 Recordati / Orphan Europe
- 10 Vertex

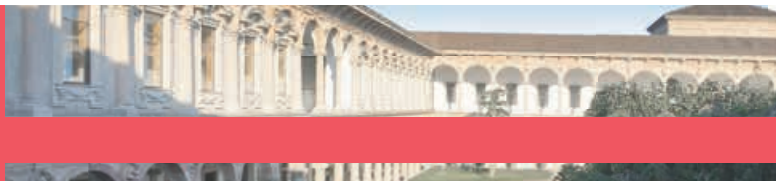


Please note that the Organising Secretariat has the right to modify the current layout, also on site for Security reasons.
Each booth is equipped with 1 table, 2 chairs, 1 paper bin and 1 electrical plug

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Adult Cystic Fibrosis



organising secretariat



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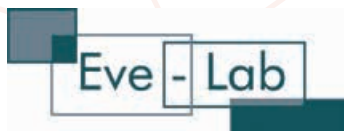


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cme accreditation



EUROPEAN CME ACCREDITATION

Please note that the 1st Adult Cystic Fibrosis International Workshop (Milan, September 5-6, 2019) has been accredited by the European Accreditation Council for Continuing Medical Education (EACCME®) with **13 European CME credits (ECMEC®s)**.



Following EBAP's evaluation, the CME activity of 1st Adult Cystic Fibrosis International Workshop (Milan, September 5-6, 2019) has been accredited by EBAP with **12 European CME credits covering the whole program**.

ITALIAN CME ACCREDITATION

The workshop has been accredited for Italian CME – E.C.M. thanks to the cooperation with the Italian CME Provider EVE-LAB (Agenas Provider n. 5306).



Italian CME Accreditation number: 5306 - 267094

Number of Italian CME Credits granted: 9,8

Accredited Professions: Medical Doctor, Nurse, Paediatric Nurse, Physiotherapist, Bio-Lab Technician

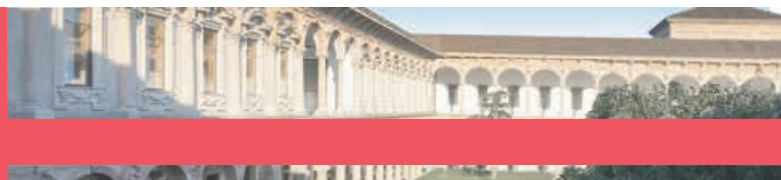
Medical Doctor Specialties: Allergology and Immunology, Internal Medicine, ENT, Respiratory Medicine, Gastroenterology, Paediatrics, Radiology

specialties

The congress programme has been designed to fulfill the educational and updating needs of professionals operating in the following specialties:

Allergology
Immunology
Internal Medicine
Otolaryngology
Respiratory Medicine
Gastroenterology

Paediatrics
Physiotherapist
Radiology
Nurses
Paediatric Nurses
Bio-Lab Technicians



scientific programme

September 5th, 2019

12.00 – 12.30 **Registration**

12.30 – 13.00 **Welcome address**
F. Blasi (Italy)

RISING STAR SESSION

The Rising Star Session hosts the presentation of two young researchers, who are going to present a scientific paper, selected by the Scientific Committee throughout a "blinded scoring process". The Call for Rising Stars has been open from October 2018 to June 2019, to all the young clinicians and researchers under 42, which have an original scientific work to be presented.

Chairs: F. Blasi, S. Aliberti, C. Colombo, S. Elborn, P. Palange, V. Raia, M. Shteinberg

13.00-13.15 **Lung Transplantation for Cystic Fibrosis: the Milan experience**
M. Contarini (Italy)

13.15 – 13.30 **Q&A and Advising time**

13.30 – 13.45 **Forskolin-induced swelling of intestinal organoids correlates with disease severity in adults with cystic fibrosis and homozygous F508del mutations**
K. de Winter - de Groot (The Netherlands)

13.45 – 14.00 **Q&A and Advising time**

SESSION 1: LIVING WITH CYSTIC FIBROSIS

Chairs: P. Palange (Italy) – J. Davies (UK)

14.00-14.30 **How I live with my Cystic Fibrosis**
J. Maetz (France)

14.30 – 15.00 **How I live as Cystic Fibrosis patient's care giver**
S. Ceri (Italy)

15.00 – 15.30 **DISCUSSION**
A. Gramegna (Italy)

15.30 – 16.00 **COFFEE BREAK & STROLLING POSTER SESSION***

SESSION 2: FROM EPIDEMIOLOGY TO GUIDELINES

Chairs: F. Blasi (Italy) – V. Raia (Italy)

16.00 – 16.30 **(Re) discovering mucoactive drugs in CF**
C. Castellani (Italy)

16.30 – 17.00 **The epidemiologic panorama of Cystic Fibrosis**
P. R. Burgel (France)

17.00 – 17.30 **The ERS/ECFS and NICE updated guidelines on the care of adults with cystic fibrosis**
S. Elborn (UK)

17.30-18.00 **DISCUSSION**
J. Altenburg (The Netherlands)

* For details related with Poster Session see p.17

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scientific programme

September 5th, 2019

18.00 – 19.00	PLENARY SYMPOSIUM
	Pushing the boundaries for Pseudomonas Aeruginosa management in Cystic Fibrosis* <i>Chair: C. S. Haworth (UK)</i>
	Welcome & Introduction <i>C. S. Haworth (UK)</i>
	Alternate or combination antibiotic regimens? Share your experience for the best patient outcomes <i>S. Elborn (UK)</i>
	Improving disease outcome by improving treatment adherence <i>P. R. Burgel (France)</i>
18.00 – 18:05	
18:05 – 18:40	
18:40 – 18:55	
18:55 – 19:00	Closing Remarks <i>C. S. Haworth (UK)</i>

19.00 – 19.30 KEY NOTE LECTURE 1*

Chair: S. Elborn (UK)

19.00 – 19.30	New treatments for Cystic Fibrosis <i>J. Davies (UK)</i>
19.30-21.00	WELCOME RECEPTION

**the session identified by "*" are not part of the CME Program, both for Italian CME and UEMS/EBAP Procedure. Participants cannot claim credits for these specific sessions*



scientific programme

September 6th, 2019

KEY NOTE LECTURE 2

Chair: V. Raia (Italy)

8.30-9.00	Transition or not transition S. Madge (UK)
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SESSION 3: MANAGEMENT OF INFECTIONS

Chairs: S. Aliberti (Italy) – M. Shteinberg (Israel)

9.00 - 9.30	Adults are different B. Plant (Ireland)
9.30 - 10.00	Inhaled antibiotics C. Schwarz (Germany)
10.00 - 10.30	Macrolides and anti-inflammatories G. Taccetti (Italy)
10.30 - 11.00	DISCUSSION J. Altenburg (The Netherlands) – A. Gramegna (Italy)

11.00-11.30	COFFEE BREAK & STROLLING POSTER SESSION*
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11.30 – 12.10 LECTURE & DEBATE

Chair: C. Schwarz (Germany)

11.30 -12.10	Continuous alternate inhaled antibiotic therapy: escalation of care or early preventive approach?" – Open Debate S. Elborn (UK)
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KEY NOTE LECTURE 3

Chair: B. Assael (Italy)

12.10 – 12.40	The Management of NTM in CF C. S. Haworth (UK)
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KEY NOTE LECTURE 4

Chair: F. Blasi (Italy)

12.40 -13.10	Mycobiome in Cystic Fibrosis and beyond: a clinical perspective S. H. Chotirmall (Singapore)
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13.10-14.30	LUNCH
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* For details related with Poster Session see p.21

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scientific programme

September 6th, 2019

SESSION 4: SPECIFIC ADULT ISSUES

Chairs: C. Colombo (Italy) – V. Makhmutova (Russian Federation)

14.30 – 15.00	Making the difficult diagnosis <i>N. J. Simmonds (UK)</i>
15.00 – 15.30	Cancer prevention: when, how, for whom? <i>P. Maisonneuve (Italy)</i>
15.30 – 16.00	Lung transplantation in CF: indications and contraindications <i>C. Benden (Switzerland)</i>
16.00 – 16.30	Management of pulmonary complications <i>L. Dupont (Belgium)</i>
16.30 – 17.00	DISCUSSION <i>L. Morlacchi (Italy)</i>
17.00 – 17.30	COFFEE BREAK

SESSION 5: LIVE AND DIE WITH CYSTIC FIBROSIS

Chairs: F. Blasi (Italy) – S. Elborn (UK)

17.30 – 18.00	Pregnancy: when and how <i>M. Shteinberg (Israel)</i>
18.00 – 18.30	End of life management <i>J. Wood (UK)</i>
18.30 – 19.15	PANEL DISCUSSION <i>Chaired by: F. Cathcart (UK)</i>

19.15 – 19.30 CLOSING REMARKS AND POSTERS AWARDS

S. Aliberti, F. Blasi, C. Colombo, S. Elborn, P. Palange, V. Raia, M. Shteinberg



posters sessions' schedule

Please note that each group posters are divided by topics and listed by first author's last name alphabetical order.

We kindly ask to poster presenters to stay next to their own poster for the whole duration of the poster session, in order to discuss the scientific paper with chairmen and the Workshop participants.

The presenters have been underlined in the authors's list.

POSTERS Sessions' SCHEDULE

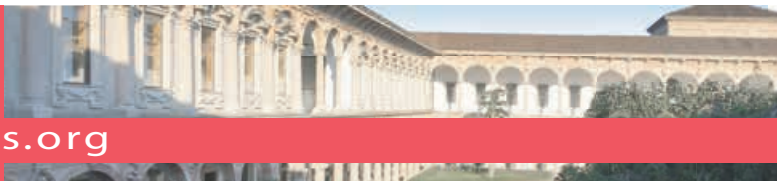
POSTER SESSION 1 September 5th, 2019 from 15:30 to 16:00

- **Group 1:** Cystic Fibrosis Epidemiology / NTM How to deal with
- **Group 2:** Lung Transplant / ERS/ECFS Guidelines in adult patients / Transition / End of life management

POSTER SESSION 2 September 6th, 2019 from 11:00 to 11:30

- **Group 1:** Management of Infections / New Treatments for Cystic Fibrosis
- **Group 2:** Patients life management / Pregnancy in Cystic Fibrosis patients

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Thursday, September 5th, 2019

15:30 - 16:00 Coffee Break & Poster Area

POSTER SESSION 1 - Group 1

Cystic Fibrosis Epidemiology
NTM How to deal with

Chairmen: S. Aliberti (Italy)
M. Shteinberg (Israel)

[1.1.1] Genetic and serum screening for alpha-1-antitrypsin deficiency in adult patients with cystic fibrosis: a single centre experience.

F. Amati¹; A. Gramegna¹; M. Contarini¹; M. Pappalettera¹; M. Seia²; L. Porcaro²; I. Ferrarotti³; A. Corsico³; S. Aliberti¹; F. Blasi¹

¹Department of Pathophysiology and Transplantation, University of Milan; Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center. Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; ²Medical Genetics Laboratory, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; ³Center for Diagnosis of Inherited Alpha1-antitrypsin Deficiency, Dept of Internal Medicine and Therapeutics, Pneumology Unit IRCCS San Matteo Hospital Foundation, University of Pavia, Pavia, Italy

[1.1.2] Immunological screening in cystic fibrosis patients revealed an unexpected high prevalence of immunodeficiencies.

F. Amati¹; A. Gramegna¹; M. Contarini¹; M. Pappalettera¹; B. Vigone²; S. Aliberti¹; F. Blasi¹

¹Department of Pathophysiology and Transplantation, University of Milan; Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center. Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; ²Scleroderma Unit, Referral Center for Systemic Autoimmune Diseases, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico di Milano, Milan, Italy

[1.1.3] Association of low BMI and fast decline of FEV1 with worse outcome in adults with CF: surveillance from a single adult centre in Slovenia

M. Badovinac¹; D. Lestan¹; H. Matevz¹; M. Turel¹; I. Kos¹; B. Salobir¹

¹UKC Ljubljana, Ljubljana, Slovenia

[1.1.4] Clinical Expression of Cystic Fibrosis patients carrying the I1234V mutation

B. E. Bar Aluma¹; D. Vilozni¹; I. Sarouk¹; Y. Bezael¹; A. Dagan¹; S. Keler¹; M. Ashkenazi¹; O. Efrati¹

¹Sackler Medical School, Tel Aviv University, Israel

[1.1.5] Clinical characteristics and disease severity of adults with cystic fibrosis and at least one residual function mutation: a multicentric study.

M. Contarini¹; A. Gramegna¹; D. Savi^{2,3}; F. Majo³; F. Ciciriello³; S. Schiavetto²; V. Lucidi³; P. Palange²; S. Aliberti¹; F. Blasi¹

¹Department of Pathophysiology and Transplantation, University of Milan; Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center. Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; ²Department of Public Health and Infectious Diseases, Adult Cystic Fibrosis Center, Sapienza University of Rome, Rome, Italy; ³Cystic Fibrosis Unit, Bambino Gesù Children's Hospital, Rome, Italy



[1.1.6] Long-term outcomes and clinical worsening in cystic fibrosis patients with at least one residual function mutation: a multicentric study.

A. Gramegna¹; M. Contarini¹; F. Majo²; D. Savi^{2,3}; F. Ciciriello²; S. Schiavetto³; P. Palange³; V. Lucidi²; S. Aliberti¹; F. Blasi¹

¹Department of Pathophysiology and Transplantation, University of Milan; Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center. Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; ²Cystic Fibrosis Unit, Bambino Gesù Children's Hospital, Rome, Italy; ³Department of Public Health and Infectious Diseases, Adult Cystic Fibrosis Center, Sapienza University of Rome, Rome, Italy

[1.1.7] Influence of genetic factors over low bone density in patients with cystic fibrosis in R. North Macedonia

T. Jakjovska¹

¹Institute for pulmonary diseases in children, Skopje, Macedonia

[1.1.8] Biofilms of Mycobacterium abscessus complex can be sensitized to antibiotics by disaggregation and oxygenation

M. Kolpen¹; P. Østrup Jensen^{1,2,3}; T. Qvist⁴; K. Nørskov Kragh^{1,3}; C. Ravnholt¹; B. Gabriel Fritz³; U. Rydahl Johansen¹; T. Bjarnsholt^{1,3}; N. Høiby^{1,3}

¹Department of Clinical Microbiology, Rigshospitalet, 2100 Copenhagen, Denmark; ²Institute for inflammation Research, Center for Rheumatology and Spine Diseases, Copenhagen University Hospital, Rigshospitalet, 2100 Copenhagen, Denmark; ³Costerton Biofilm Center, Institute of Immunology and Microbiology, Faculty of Health and Medical Sciences, University of Copenhagen, 2200 Copenhagen, Denmark; ⁴Copenhagen CF center, Department of Infectious Diseases, Rigshospitalet, 2100, Denmark

[1.1.9] Cystic Fibrosis diagnosis in adult life in Italy. Data from the Italian registry

R. Padoan¹; S. Quattrucci²; V. Carnovale³; M. Salvatore⁴; B. Giordani⁵

¹Italian Cystic Fibrosis Registry, Roma, Italy; ²Dipartimento Pediatria, Università Sapienza, Roma, Italy; ³CRR Fibrosi Cistica dell'Adulto, AOU Federico II, Università di Napoli, Napoli, Italy; ⁴National Center for Rare Diseases, Istituto Superiore di Sanità, Roma, Italy; ⁵Lega Italiana Fibrosi Cistica - onlus, Roma, Italy

[1.1.10] Air pollution exposure is associated with lung function decrease in adults with cystic fibrosis

V. Vinnat¹; I. Annesi-Maesano¹; P. Regis Burgel²

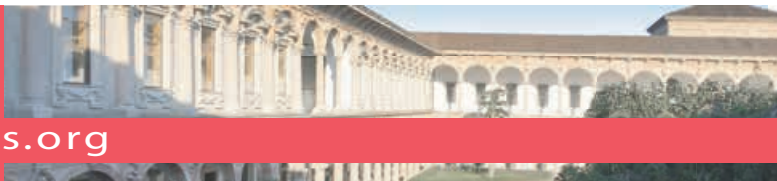
¹EPAR, IPLESP, INSERM and Sorbonne Université, Medical School Saint-Antoine, Paris, France; ²Service de pneumologie, hôpital Cochin, Paris, France

[1.1.11] Epidemiology of European adults with Cystic Fibrosis

A. Zolin¹; L. Naehrlich²; A. Fox³; M. Krasynk⁴; A. Orenti¹; J. van Rens⁵

¹University of Milan, Department of Clinical Science and Community Health, Milano, Italy; ²Justus-Liebig-University, Department of Pediatrics, Giessen, Germany; ³European Cystic Fibrosis Patient Registry, Verona, Italy; ⁴European Cystic Fibrosis Patient Registry, Lviv, Ukraine; ⁵European Cystic Fibrosis Patient Registry, Leuven, Belgium

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Thursday, September 5th, 2019

POSTER SESSION 1 - Group 2

Lung Transplant

ERS/ECFS Guidelines in adult patients

Transition

End of life management

Chairmen: C. Colombo (Italy)

P. Palange (Italy)

15:30 - 16:00 Coffee Break & Poster Area



[1.2.1] Lung Transplantation for Cystic Fibrosis: the Milan experience.

M. Contarini¹; L. C. Morlacchi¹; V. Rossetti¹; L. Rosso²; M. Nosotti²; M. Pappalettera¹; P. Tarsia¹

¹Respiratory Unit and Cystic Fibrosis Adult Centre, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico di Milano; Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Milan, Italy; ²U.O. Chirurgia Toracica e dei Trapianti di Polmone, IRCCS Fondazione Ca' Granda Ospedale Maggiore Policlinico di Milano; Università degli Studi di Milano, Milano, Italy, Milano, Italy

[1.2.2] Combined Kidney-Lung Transplant under venovenous-ECMO in a Cystic Fibrosis patient

C. Dantas¹; A. S. Santos¹; L. Semedo¹; P. Calvino²; F. Remedio³; F. Nolasco³; J. Cardoso¹; J. Fragata²

¹Pulmonology Department - CHULC, Lisbon, Portugal; ²Cardiothoracic Surgery Department - CHULC, Lisbon, Portugal; ³Nephrology Department - CHULC, Lisbon, Portugal



[1.2.3] Forskolin-induced swelling of intestinal organoids correlates with disease severity in adults with cystic fibrosis and homozygous F508 del mutations

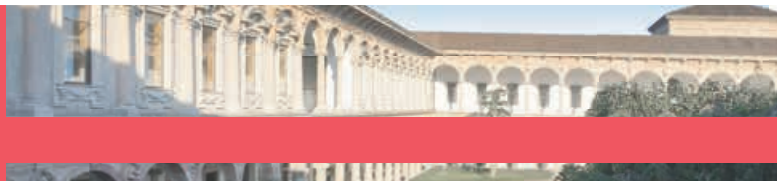
K. de Winter - de Groot¹; G. Berkers¹; R. van der Meer²; A. Vonk^{1,3}; S. Michel¹; E. Kruisselbrink^{1,3}; R. Vries⁴; H. Clevers⁵; F. Vleggaar⁶; S. Elias⁷; H. Heijerman^{2,8}; K. van der Ent¹; J. Beekman^{1,3}

¹Cystic Fibrosis Center, University Medical Center Utrecht, Utrecht, Netherlands; ²Cystic Fibrosis Center, Haga Teaching Hospital, The Hague, Netherlands; ³Department of Regenerative Medicine, University Medical Center Utrecht, Utrecht, Netherlands; ⁴Foundation Hubrecht Organoid Technology (HUB), Utrecht, Netherlands; ⁵Hubrecht Institute for Developmental Biology and Stem Cell Research, University Medical Center Utrecht, Utrecht, Netherlands; ⁶Department of Gastroenterology & Hepatology, University Medical Center Utrecht, Utrecht, Netherlands; ⁷Julius Center for Health Sciences and Primary Care, University Medical Center Utrecht, Utrecht, Netherlands; ⁸Department of Pulmonology, University Medical Center Utrecht, Utrecht, Netherlands

[1.2.4] Swiss recommendations for adult cystic fibrosis care: an open-access, point-of-care resource for adult CF centers

A. Koutsokera¹; C. Benden²; J. M. Fellrath³; R. Fischer Biner⁴; T. Geiser⁵; M. Hofer⁶; R. Kleiner⁷; L. Nicod¹; J. Plojoux⁸; A. Sauty³

¹Lausanne University Hospital, Lausanne, Switzerland; ²University Hospital Zurich, Zurich, Switzerland; ³Neuchâtel Hospital, Neuchâtel, Switzerland; ⁴Bern Lindenhofspital, Bern, Switzerland; ⁵Bern University Hospital, Bern, Switzerland; ⁶Winterthur Hospital, Winterthur, Switzerland; ⁷St. Gallen Hospital, St. Gallen, Switzerland; ⁸Geneva University Hospital, Geneva, Switzerland



[1.2.5] A first experience with a new patient-centered protocol for transition from pediatric to adult care for patients with cystic fibrosis

R. Lub¹ ; N. Gilst V.¹; N. Rutjes¹; M. Verkleij¹; H. Eeman¹; V. D. L. Schaaf¹; V. M. Brederode¹; J. Altenburg¹

¹Amsterdam MCD research group, dept of Pulmonary Diseases and Pediatrics of Amsterdam UMC, Amsterdam, Netherlands

[1.2.6] Adult CF patient, treating complications and management of the end life story

S. Momchilovikj¹ ; T. Jakjovska¹; I. Arnaudova-Danevska¹; E. Gjinovska-Tasevska¹; A. Andonovski¹

¹Institute for pulmonary diseases, Skopje, Macedonia

[1.2.7] Adherence to medical regimens after lung transplantation increased using motivational interviewing and questionnaires

U. Skogeland¹ ; I. de Monestrol¹; K. Cedermark¹

¹Stockholm CF-center, Karolinska University Hospital, Karolinska Institutet, Stockholm, Sweden

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Friday, September 6th, 2019

11:00 - 11:30 Coffee Break & Poster Area

POSTER SESSION 2 - Group 1
Management of Infections
New Treatments for Cystic Fibrosis

Chairmen: F. Blasi (Italy)
V. Raia (Italy)

[2.1.1] Fungal prevalence and diversity in Stockholm CF center, a 16 years retrospective study
M. Al Shakrichi¹; L. Klingspor²; L. Hjelte¹; K. Ceder¹; P. Bergman³; I. de Monestrol¹

¹Stockholm Cystic Fibrosis Centre, Karolinska University Hospital Huddinge, Karolinska Institutet, Stockholm, Sweden;

²Department of Laboratory Medicine, Division of Clinical Microbiology, Karolinska Institutet, Stockholm, Sweden;

³Infection Disease Clinic, The immunodeficiency Unit, Karolinska University Hospital Huddinge, Stockholm, Sweden

[2.1.2] Use of Ivacaftor in Cystic Fibrosis patients with residual function CFTR mutations

I. Bonato¹; S. Santaniello¹; F. Cresta¹; R. Casciaro¹; C. Castellani¹

¹Cystic Fibrosis Center, IRCCS Giannina Gaslini Institute, Genova, Italy

[2.1.3] The impact of Lumacaftor/Ivacaftor treatment on airways' microbiology

E. Bourgani¹; C. Kosti¹; E. Stagaki¹; T. Panagea²; F. Diamantea¹

¹Adult Cystic Fibrosis Unit, Sismanoglio General Hospital, Athens, Greece; ²Department of Clinical Microbiology, Sismanoglio General Hospital, Athens, Greece

[2.1.4] Role for cytokines on mucus production and attachment to the airway

M. Giorgetti¹; A. Ermund¹; G. Hansson¹

¹Department of Medical Biochemistry, University of Göteborg, Göteborg, Sweden

[2.1.5] Multidrug resistant new, previously unculturable bacterium associated with adult cystic fibrosis Chryseobacterium mucoviscidosis VT16-26

C. Kardava¹; G. Tetz¹; M. Vecherkovskaya¹; T. Gembitskaia²; V. Tetz¹

¹Department of Microbiology and Virology, Pavlov First Saint Petersburg State Medical University, Saint Petersburg, Russian Federation; ²Pulmonology Research Institute, Pavlov First Saint Petersburg State Medical University, Saint Petersburg, Russian Federation

[2.1.6] Lumacaftor/Ivacaftor combination in Cystic Fibrosis (CF) patients: real-life data from Verona CF Center

F. Lucca¹; G. Cucchetto²; E. Spinelli²; E. Pintani²; S. Volpi²

¹Pediatrics School, University of Verona, Verona, Italy; ²CF Center, Verona Hospital, Verona, Italy



[2.1.7] Antimicrobial susceptibility of bacteria isolated from adult patients with cystic fibrosis treated in the Institute for Pulmonary Diseases of Vojvodina Sremska Kamenica, Serbia

D. Skrbic¹; A. Trudic^{1,2}; M. Djuric^{1,2}; T. Kurucin¹; S. Hromis^{1,2}; M. Hadnadjev¹; B. Tusek¹; D. Povazan^{1,2}

¹Institute for Pulmonary Diseases of Vojvodina, Put Doktora Goldmana 4, 21204 Sremska Kamenica, Novi Sad, Serbia; ²University of Novi Sad, Faculty of Medicine, Hajduk Veljkova ³, 21000 Novi Sad, Novi Sad, Serbia

[2.1.8] The effect of Lumacaftor/Ivacaftor on the severity of CF pulmonary exacerbations in patients with F508 del homozygous CF

K. Yaacoby-Bianu¹; S. Nadeem²; S. Nili³; Z. Schnapp⁴; L. Galit¹; M. Shteinberg^{2,5}

¹Pediatric Pulmonology Unit and CF Center, Carmel Medical Center, Haifa, Israel; ²B. Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel; ³Department of Epidemiology and community health, Carmel Medical Center, Haifa, Israel; ⁴Department of Pediatrics, Carmel Medical Center, Haifa, Israel; ⁵Pulmonology Institute and CF Center, Carmel Medical Center, Haifa, Israel

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Friday, September 6th, 2019

POSTER SESSION 2 - Group 2
Patients Life Management
Pregnancy in Cystic Fibrosis patients

Chairmen: C. Colombo (Italy)
S. Elborn (UK)

11:00 - 11:30 Coffee Break & Poster Area

[2.2.1] Doctoral thesis: Adults with cystic fibrosis: mental health and patient experiences of the CF treatment
L. Backström Eriksson¹

¹Karolinska Institutet, Division of psychology, Stockholm, Sweden

[2.2.2] Swedish “middle aged”, non-transplanted CF-patients’ experiences of their daily multifactorial treatment: a qualitative study

L. Backström Eriksson^{1,2}; L. Hjelte^{1,3}; B. Melin²; K. Sorjonen²; M. Lundberg⁴

¹Karolinska University Hospital, Stockholm CF-center, Stockholm, Sweden; ²Karolinska Institutet, Division of psychology, Stockholm, Sweden; ³Karolinska Institutet, Division of Paediatrics, Stockholm, Sweden; ⁴University of Gothenburg, Department of Health and Rehabilitation, Gothenburg, Sweden

[2.2.3] Perinatal outcomes in Cystic Fibrosis women. Data from the Italian Cystic Fibrosis Registry (ICFR)
R. Padoan¹; A. Amato²; G. Barbara²; F. Majo³; S. Quattrucci⁴; M. Salvatore⁵; D. Salvatore⁶; V. Carnovale⁷

¹Italian Cystic Fibrosis Registry, Rome, Italy; ²Lega Italiana Fibrosi Cistica onlus, Rome, Italy; ³CF Center, Bambino Gesù Hospital, Rome, Italy; ⁴Pediatric Department Sapienza University, Rome, Italy; ⁵National Center for Rare Diseases, Istituto Superiore di Sanità, Rome, Italy; ⁶CF Center, Hospital San Carlo, Potenza, Italy; ⁷CRR Fibrosi Cistica dell'Adulto, AOU Federico II, Università di Napoli, Napoli, Italy

[2.2.4] Survivors Against All Odds

K. Radwan¹; Å. Silfverplatz¹; A. Jarblad¹; I. de Monestrol²; A. B. Brucefors³

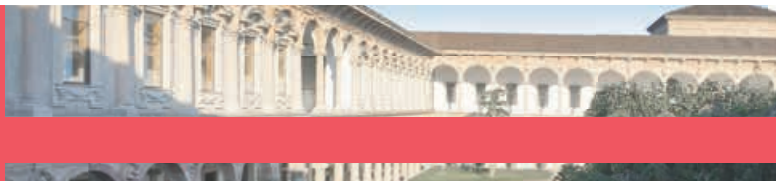
¹Swedish Cystic Fibrosis Association, RfCF, Uppsala, Sweden; ²Stockholm CF Centre, Karolinska University Hospital, Stockholm, Sweden; ³Retired, Previously Stockholm CF Centre, Karolinska University Hospital, Stockholm, Sweden

[2.2.5] Accelerated progression of Cystic Fibrosis lung disease during pregnancy with fatal outcome
K. Templeton¹; S. Thomson¹; E. Ross¹; J. Brennand²; G. MacGregor¹; S. Bickenell¹

¹West of Scotland Cystic Fibrosis Service, Glasgow, United Kingdom; ²Department of Obstetrics, Queen Elizabeth University Hospital, Glasgow, United Kingdom

[2.2.6] Pregnancy outcomes in women with Cystic Fibrosis: 10 year experience from the west of Scotland
K. Templeton¹; E. Ross¹; S. Thomson¹; J. Brennand²; S. Bicknell¹; G. Macgregor¹

¹West of Scotland Cystic Fibrosis Service, Glasgow, United Kingdom; ²Department of Obstetrics, Queen Elizabeth University Hospital, Glasgow, United Kingdom



endorsements

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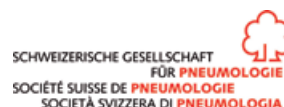
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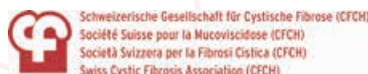
PATIENTS' ASSOCIATIONS



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contre la Mucoviscidose



LIFC
Lega Italiana
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workshop general information

ADMISSION

The participant's name badge will be provided at the registration desk. All participants are requested to wear the badge throughout the congress. Only badge holders will be admitted to the appropriate sessions, exhibition and social events.

PRESENCE CONTROL

Effective attendance is checked through badge scanning when going in and coming out of the plenary room.

CERTIFICATE OF ATTENDANCE

The certificate of attendance will be downloadable from the website www.adultcysticfibrosis.org using the personal log-in credentials sent by e-mail, starting from September 11th, 2019.

Important note: Certificate of attendance would be delivered only to registered participants who attend at least 6 hours of congress sessions.

For Sponsored Groups: Please note that in order to send the certificate of attendance to each participant, we will need the personal e-mail.

LANGUAGE

The official language of the Congress is English. No simultaneous translation will be provided.

SECRETARIAT

The secretariat desk will open on Thursday, September 5th, 2019 at 12.00, and stay open throughout the Congress.

INSURANCE

The Congress organizers cannot accept liability for personal injuries sustained, or for loss or damage to property belonging to Congress participants, either during or as a result of the Congress. Registration does not include insurance. It is strongly recommended that you take an insurance policy of your choice as you register for the Congress and book your travel.

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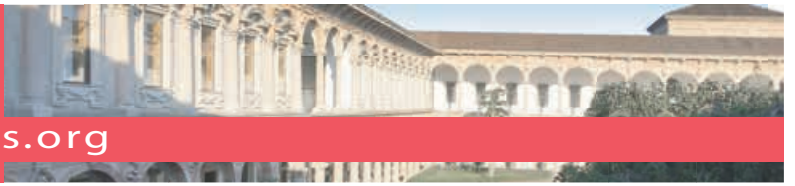
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I-neb AAD, the smart device:

Adaptive Aerosol Delivery (AAD) + Electronic Adherence Monitoring

Each patient is different! I-neb AAD adapts to all patients' unique breathing characteristics:

- Breath activated: it pulses aerosol only during inhalation¹
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- I-neb delivers precise and reproducible doses of aerosol¹
- I-neb in TIM mode guides the patients to slow and deep inhalations to improve lung deposition¹



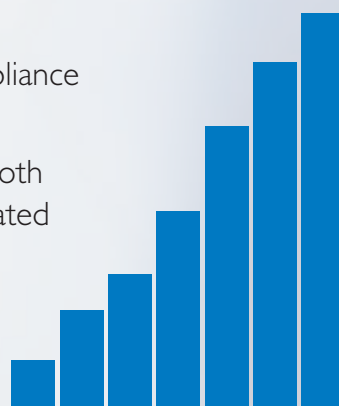
Breaking down barriers to medication adherence

Paediatric Respiratory Reviews:

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I-neb: True Adherence

- I-neb records data on patient's compliance to everyday treatment regimens¹
- The data can be easily analyzed by both clinicians and patients through dedicated software on a PC¹
- Electronic adherence monitoring can help to improve patient's adherence¹



Reference:

1. John Denyer et al., The Adaptive Aerosol Delivery (AAD) Technology: Past, Present, and Future, journal of aerosol medicine and pulmonary drug delivery. Volume 23, Supplement 1, 2010
2. Wildman MJ, Hoo ZH. Moving cystic fibrosis care from rescue to prevention by embedding adherence measurement in routine care. Paediatr Respir Rev 2014; 15(1): 16-18

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