

Milan
5-6 September
2019

Università degli Studi di Milano



1st Adult Cystic Fibrosis International Workshop



Adult Cystic Fibrosis

POSTER SESSIONS

12

EBAP
credits
GRANTED

13

EUROPEAN CME
credits (ECMEC®S)
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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

Organised by:



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. Shtenberg (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. Vilozeni¹; I. Sarouk¹; Y. Bezalel¹; 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

Organised by:



Thursday, September 5th, 2019

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

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)



[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, 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. Calvinho²; 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. Shtenberg^{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

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

POSTER SESSION 2 - Group 2

Patients Life Management

Pregnancy in Cystic Fibrosis patients

Chairmen: C. Colombo (Italy)

S.Elborn (UK)

[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