Scientific Programme

Wednesday, 06 June 2018

Meetings and Courses
09:00 - 17:00
Room 15

An introductory course in cognitive behavioural therapy and interpersonal therapy**
** Extra registration required
Alexandra Klein Rafaeli (Tel Aviv, Israel)

- CBT principles: assessment and formulation, session structure, cognitive restructuring, exposure experiments, transdiagnostic treatment goals, monitoring and homework
- The third wave of CBT: introduction to mindfulness
- Coffee Break
- What is IPT and how is it similar to/different from CBT?
- Lunchbreak
- How to conduct the interpersonal inventory
- Coffee Break
- IPT Case Formulation: identifying interpersonal problem areas  IPT Techniques: communication analysis and roleplaying  Termination Phase and Conclusion (CBT and IPT)

Meetings and Courses
10:00 - 16:40
Amphitheatre

The treatment of adults with CF - an in-depth course

Wednesday 6 June - All day

In many countries, there are now more adults than children with CF. Therefore, this course is specifically designed for pulmonologists and invites them to deepen their knowledge on the treatment of adults with CF.

Carsten Schwarz (Berlin, Germany)
Stuart Elborn (London, United Kingdom)

- Introduction and adult CF epidemiology
  Pierre-Régis Burgel (Paris, France)
- Gram-negative infections and their treatment
  Lieven Dupont (Leuven, Belgium)
- ‘Other’ pathogens and their treatment
  Carsten Schwarz (Berlin, Germany)
- Pulmonary complications and their treatment
  Nicholas Simmonds (London, United Kingdom)
- Lunch break
Scientific Programme

**Comorbidities in adults with CF**
Stuart Elborn (London, United Kingdom)

**Standards of Care / Segregation / Hygiene / Multidisciplinary teams / Quality control**
Daniel Peckham (Leeds, United Kingdom)

**Coffee Break**

**Pipeline of treatments correcting the basic CF defect**
Stuart Elborn (London, United Kingdom)

**Discussion on the need for and challenges of adult CF clinics in the Balkans**
Predrag Minic (Belgrade, Serbia)

**Closing remarks**

**Physiotherapy Case Presentations**

**Opening Plenary**
18:30 - 20:00

**Opening Plenary**
Kris De Boeck (Leuven, Belgium)
Predrag Minic (Belgrade, Serbia)

**Welcome Address**
Predrag Minic (Belgrade, Serbia)

**Presentation of the ECFS Award**

**ECFS Award Lecture**
Susan Madge (London, United Kingdom)

**Clinical trials from an ethical perspective**
Suzanne van de Vathorst (Rotterdam, Netherlands)

**Presentation of the Gerd Döring Award**

**Presentation of the CFE Advocacy Award**

**Welcome Reception**
20:00 - 21:30
Thursday, 07 June 2018

Satellite Symposium
07:15 - 08:15

Satellite Symposium
Please find the detailed program of the Satellite Symposia here.

Symposium
08:30 - 10:00

S01, Microbiome directed treatment

Upon completion of this session participants should be able to:
- Have an overview of the lung and gut microbiome in CF
- Understand the impact of antibiotic treatment on the lung and gut microbiome
- Learn if microbiome directed antibiotic therapy is more effective than conventional microbiology in treating pulmonary exacerbations in CF

Michael Tunney (Belfast, United Kingdom)
Barry Plant (Cork, Ireland)

<table>
<thead>
<tr>
<th>Session Title</th>
<th>Speaker(s)</th>
<th>Time</th>
</tr>
</thead>
<tbody>
<tr>
<td>The CF microbiome - where are we now?</td>
<td>Debby Bogaert (Edinburgh, United Kingdom)</td>
<td>08:30 - 08:52</td>
</tr>
<tr>
<td>Microbiome directed antibiotic treatment of pulmonary exacerbations - results of an international randomised controlled trial</td>
<td>Barry Plant (Cork, Ireland)</td>
<td>08:52 - 09:14</td>
</tr>
<tr>
<td>The lung/gut axis - longitudinal microbiome</td>
<td>Gisli Einarsson (Belfast, United Kingdom)</td>
<td>09:14 - 09:36</td>
</tr>
<tr>
<td>Immune response in a mouse model of chronic infection</td>
<td>Pierre-Régis Burgel (Paris, France)</td>
<td>09:36 - 10:00</td>
</tr>
</tbody>
</table>

Symposium
08:30 - 10:00

S02, CF centre care of the future

Upon completion of this session participants should be able to:
- Learn how to reduce the risk of cross infection in CF centres of the future
- Understand how new technologies will enable better monitoring of patients and potentially earlier intervention
- Understand how trans-border networks could enhance clinical care

Andres Floto (Cambridge, United Kingdom)
Hannah Blau (Petah Tikva, Israel)

<table>
<thead>
<tr>
<th>Session Title</th>
<th>Speaker(s)</th>
<th>Time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optimising new CF centre design from an infection, prevention and control perspective</td>
<td>Lisa Saiman (New York, United States)</td>
<td>08:30 - 08:52</td>
</tr>
</tbody>
</table>
Scientific Programme

**How remote monitoring and biosensor technologies may change CF management?**
Jane Davies (London, United Kingdom)
08:52 - 09:14

**Use of machine learning to predict pulmonary exacerbations**
Andres Floto (Cambridge, United Kingdom)
09:14 - 09:36

**Trans-border advice: its place in CF care and the role of the European Reference Network to implement it**
Kors Van der Ent (Utrecht, Netherlands)
09:36 - 10:00

**S03, Instructions for a wise use of CF genetics**

The objectives of the session are to:
- Summarise current knowledge on the interactions between the genotype and phenotype
- Explain the new terminology of theratypes
- Appraise home genetic testing for CF

Dragica Radojković (Belgrade, Serbia)
Milan Jr Macek (Prague, Czech Republic)

**European quality control for CFTR genetic analysis - the results**
Els Dequeker (Leuven, Belgium)
08:30 - 08:52

**Direct to consumer genetic tests: what is their safety and efficacy? How to interpret their results?**
Milan Jr Macek (Prague, Czech Republic)
08:52 - 09:14

**Theratypes: what are they? How can they help treating patients?**
Margarida Amaral (Lisbon, Portugal)
09:14 - 09:36

**Update on CF modifier genes**
Harriet Corvol (Paris, France)
09:36 - 10:00

**S04, Start of life to end of life: living with uncertainty**

Upon completion of this session participants should be able to:
- Summarise the complexities associated with newborn screening in CF and consider how these are communicated to carers and colleagues
- Consider how teams can prepare for the changing needs of patients with CF in the 21st century
- Discuss how CFTR-RD is managed in adult services and critically reflect on learning from these cases to help guide the care of CFSPID-CRMD

Anne Munck (Paris, France)
Urszula Borawska - Kowalczyk (Warsaw, Poland)

**Start of life discussions - what do these genes mean? - communication complexity**
Colin Wallis (London, United Kingdom)
08:30 - 08:52
41st European Cystic Fibrosis Conference,
07 - 10 June 2017,
Belgrade, Serbia

Scientific Programme

Changing (un)certainties in CF and the challenges for MD teams 08:52 - 09:14
Susan Madge (London, United Kingdom)

CFTR-RD: is this adult CFSPID? 09:14 - 09:36
Carlo Castellani (Verona, Italy)

Uncertainty for the MDT: how is CFTR-RD managed? 09:36 - 10:00
Isabelle Durieu (Lyon, France)

Symposium
08:30 - 10:00
Annex A

S05, Evolving the role of physiotherapy
Marta Kerstan (Biberstein, Switzerland)
Ruth Dentice (Sydney, Australia)

Arthropathy and arthritis 08:30 - 08:52
Elizabeth Clarke (Manchester, United Kingdom)

Coaching for success, the upper airway 08:52 - 09:14
William Poncin (Brussels, Belgium)

Incontinence 09:14 - 09:36
Sophie Ramel (Roscoff, France)

How to treat pain? 09:36 - 10:00
Dominique Hubert (Paris, France)

Symposium
08:30 - 10:00
Annex B

S06, What’s going on below the diaphragm?
Upon completion of this session participants should be able to:
- Better understand the impact of nutrition on the gut microbiome
- Learn that pancreatitis is more than pancreatitis
- Learn the impact of endocrine dysfunction of the pancreas on disease progression
- Learn the importance of recognising the micronutrient deficiency in CF

Stephanie Van Biervliet (Ghent, Belgium)
Duška Drinković-Tješić (Zagreb, Croatia)

The influence of diet on gut microbiome 08:30 - 08:52
Daniel Peckham (Leeds, United Kingdom)

Two questions on pancreatitis: when is it worth thinking of cystic fibrosis / can new therapies increase the incidence? 08:52 - 09:14
Michael Wilschanski (Jerusalem, Israel)

Pancreatic endocrine dysfunction in cystic fibrosis 09:14 - 09:36
Antoinette M. Moran (Minneapolis, United States)
41st European Cystic Fibrosis Conference, 07 - 10 June 2017, Belgrade, Serbia

**Scientific Programme**

**Recognising trace elements and vitamin deficiencies**  
Konstantinos Gerasimidis (Glasgow, United Kingdom)  
09:36 - 10:00

**Satellite Symposium**  
10:00 - 10:30  
ePoster Corners

**Satellite Workshop**  
ePoster Corner A  
Please find the detailed program of this session [here](#).

**Symposium**  
10:30 - 12:00  
Blue Hall

**S07, Mix and match! Combination CFTR modulation therapies and new trials**

Upon completion of this session participants should be able to:
- Describe approaches of CFTR modulation in different drug development programs
- Evaluate how different correctors and potentiators may impact CFTR function
- Assess effectiveness and side effects of new correctors and potentiators

Pavel Drevinek (Prague, Czech Republic)  
Silke van Koningsbruggen-Rietschel (Cologne, Germany)

- **Galapagos: new study results/development pipeline**  
  Katja Conrath (Mechelen, Belgium)  
  10:30 - 10:52

- **Flatley: new study results/development pipeline**  
  Claudia Ordonez (Charlestown, United States)  
  10:52 - 11:14

- **Proteostasis: new study results/development pipeline**  
  Geoffrey Gilmartin (Cambridge, United States)  
  11:14 - 11:36

- **Vertex: new study results/development pipeline**  
  Charlotte McKee (Boston, United States)  
  11:36 - 12:00

**Symposium**  
10:30 - 12:00  
Hall 1

**S08, How do pathogens become multiresistant?**

Upon completion of this session participants should be able to:
- Classify the key mechanisms of bacterial resistance to antibiotics and how pathogens develop or acquire resistance
- Identify antibiotic resistance rates of CF pathogens and their clinical implications
- Discuss how choosing antibiotics based on biofilm rather than conventional antimicrobial susceptibility testing could influence treatment response

Barbara Kahl (Münster, Germany)  
Eshwar Mahenthiralingam (Cardiff, United Kingdom)

- **Efflux pumps and resistance mechanisms in CF pathogens**  
  Silvia Buroni (Pavia, Italy)  
  10:30 - 10:52
Mechanisms of resistance and susceptibility in CF pathogens
Francoise van Bambeke (Brussels, Belgium) 10:52 - 11:14

Evolution of resistance
Carla Lopez Causapé (Palma de Mallorca, Spain) 11:14 - 11:36

Biofilm resistance and testing
Claus Moser (Copenhagen, Denmark) 11:36 - 12:00

Symposium
10:30 - 12:00

S09, NTM: sources, transmission and treatments

Upon completion of this session participants should be able to:
- Understand the evidence supporting Pseudomonas eradication regimens
- Learn about the natural history and treatment approaches to other difficult Gram-negative organisms
- Understand the evidence supporting MRSA eradication regimens

Charles Haworth (Cambridge, United Kingdom)
Rachel Thomson (Brisbane, Australia)

Environmental sources of NTM infection
Rachel Thomson (Brisbane, Australia) 10:30 - 10:52

Patient-to-patient transmission: whole genome sequencing update
Marianne Skov (Copenhagen, Denmark) 10:52 - 11:14

M. abscessus treatment outcomes in cystic fibrosis
Charles Haworth (Cambridge, United Kingdom) 11:14 - 11:36

Future therapies for NTM lung disease
Andres Floto (Cambridge, United Kingdom) 11:36 - 12:00

Symposium
10:30 - 12:00

S10, Best of Journal of Cystic Fibrosis and Lancet Respiratory Medicine

This session will provide a forum for attendees to interact with the authors and editors of papers published in the journals. Papers presented will be recent publications, selected by the editors, and attendees will have the opportunity to hear presentations directly from the authors and editors, and address questions to both the authors and editors. The discussion is intended to provide insights into these papers, the selection process, and how the research applies directly to the field of CF.

Scott Bell (Brisbane, Australia)
Emma Grainger (London, United Kingdom)

Welcome
Scott Bell (Brisbane, Australia) 10:30 - 10:39
41st European Cystic Fibrosis Conference,
07 - 10 June 2017,
Belgrade, Serbia

Scientific Programme

Journal of Cystic Fibrosis One-time quantitative PCR detection of Pseudomonas aeruginosa to discriminate intermittent from chronic infection in cystic fibrosis
Alexander Dalpke (Heidelberg, Germany)
Neils Hoiby (Denmark) 10:39 - 10:57

The Lancet Respiratory Medicine Article 01
Journal of Cystic Fibrosis Comparison of ex vivo and in vitro intestinal CF models to measure CFTR-dependant ion channel transport
Jeffrey Beekman (Utrecht, Netherlands)
Nicholas Simmonds (London, United Kingdom) 10:57 - 11:15

11:15 - 11:33

Journal of Cystic Fibrosis Comparison of ex vivo and in vitro intestinal CF models to measure CFTR-dependant ion channel transport
Jeffrey Beekman (Utrecht, Netherlands)
Nicholas Simmonds (London, United Kingdom) 11:15 - 11:33

The Lancet Respiratory Medicine Article 02
Conclusions
Emma Grainger (London, United Kingdom) 11:33 - 11:51

11:51 - 12:00

Symposium
10:30 - 12:00

Annex A

S11, Fertility and reproductive challenges in cystic fibrosis

Upon completion of this session participants should be able to:
- Understand the importance of preconception counselling regarding parental genetics and longevity, appropriate medication use during pregnancy, and diabetes surveillance and control
- Learn about the management of male infertility
- Appreciate the complexities of pulmonary and extra-pulmonary management during pregnancy including antibiotic strategies, airway clearance strategies, the management of haemoptysis, ventilatory failure, nutrition, diabetes, and liver disease

Malena Cohen-Cymberknoh (Jerusalem, Israel)
Susan Madge (London, United Kingdom)

Preconception planning
Hannah Blau (Petah Tikva, Israel) 10:30 - 10:52

Management of male infertility
Susan Madge (London, United Kingdom) 10:52 - 11:14

Pulmonary management during pregnancy
Malena Cohen-Cymberknoh (Jerusalem, Israel) 11:14 - 11:36

Management of extrapulmonary complications during pregnancy
Jennifer Taylor-Cousar (Denver, United States) 11:36 - 12:00
Scientific Programme

Symposium
10:30 - 12:00

S12, CFTR from structure to regulation

Upon completion of this session participants should be able to:
- Describe our present knowledge of the tridimensional structure of CFTR protein
- Discuss the importance of molecular interactions of CFTR with other proteins
- Appraise the potential use of novel structural information for the development of improved pharmacological modulators of mutant CFTR

Isabelle Callebaut (Paris, France)
Bertrand Kleizen (Utrecht, Netherlands)

<table>
<thead>
<tr>
<th>Time</th>
<th>Topic</th>
<th>Speaker(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>10:30 - 10:52</td>
<td>CFTR 3-D structure and interaction with therapeutic drugs</td>
<td>Bob Ford (Manchester, United Kingdom)</td>
</tr>
<tr>
<td>10:52 - 11:14</td>
<td>Effect of modulators on cooperative folding</td>
<td>Bertrand Kleizen (Utrecht, Netherlands)</td>
</tr>
<tr>
<td>11:14 - 11:36</td>
<td>Molecular interactions at the plasma membrane to increase CFTR function</td>
<td>Anjaparavanda P. Naren (Cincinnati, United States)</td>
</tr>
<tr>
<td>11:36 - 12:00</td>
<td>Identification of new active compounds using improved cellular assays</td>
<td>Paola Vergani (London, United Kingdom)</td>
</tr>
</tbody>
</table>

Satellite Symposium
12:30 - 14:00

Satellite Symposium

Please find the detailed program of the Satellite Symposia here.

Meet the Experts
12:45 - 13:45

Meet the Experts 1: Multidisciplinary team function and dysfunction

ePoster Corner A

Trudy Havermans (Leuven, Belgium)
Susan Madge (London, United Kingdom)
Meet the Experts
12:45 - 13:45

**Meet the Expert 2: Match the drug to the bug**

ePoster Corner B

Helle Krogh Johansen (Copenhagen, Denmark)
Giovanni Taccetti (Florence, Italy)

Meet the Experts
12:45 - 13:45

**Meet the Expert 3: Feeding behaviour in cystic fibrosis**

ePoster Corner C

Mandy Bryon (London, United Kingdom)
Jacqueline Lowdon (Leeds, United Kingdom)

ECFS Tomorrow Session
12:45 - 13:45

**Mental health Guidelines from East to West (and back). Local adaptation of the implementation of the MH guidelines**

Pavla Hodkova (Prague, Czech Republic)
Anna Georgioupolos (Boston, United States)
Diana Kadosh (Petah Tikva, Israel)
Urszula Borawska - Kowalczyk (Warsaw, Poland)

ePoster Session
14:00 - 15:00

**ePS3, New treatments: from genes to protein**

ePoster Corner C

Marcus Mall (Berlin, Germany)
Jane Davies (London, United Kingdom)

- **Introduction**
  14:00 - 14:04

- **Transduction of rhesus macaque lung following repeat dosing by adenoviral-associated virus serotype 1**
  14:04 - 14:12
  Liudmila Cebotaru (Baltimore, United States)

- **Exploratory immune assays distinguish healthy volunteer from cystic fibrosis patient cohorts and were validated in a dose escalation study of QR-010 in subjects with cystic fibrosis homozygous for the F508del CFTR mutation**
  14:12 - 14:20
  Miriam Bujny (Leiden, Netherlands)
Scientific Programme

Effects of lumacaftor-ivacaftor therapy on CFTR function in Phe508del homozygous patients with cystic fibrosis
Simon Graeber (Heidelberg, Germany)
14:20 - 14:28

Impact of CFTR modulation with ivacaftor on gut microbiota and intestinal inflammation
Chee Y. Ooi (Randwick, Australia)
14:28 - 14:36

GLPG2222 in subjects with cystic fibrosis homozygous for F508del: results from a phase II study (FLAMINGO)
Kors Van der Ent (Utrecht, Netherlands)
14:36 - 14:44

GLPG2222 in subjects with cystic fibrosis homozygous for F508del: results from a phase II study (FLAMINGO)
Kors Van der Ent (Utrecht, Netherlands)
14:36 - 14:44

GLPG2222 in subjects with cystic fibrosis homozygous for F508del: results from a phase II study (FLAMINGO)
Kors Van der Ent (Utrecht, Netherlands)
14:36 - 14:44

GLPG2222 in subjects with cystic fibrosis homozygous for F508del: results from a phase II study (FLAMINGO)
Kors Van der Ent (Utrecht, Netherlands)
14:36 - 14:44

GLPG2222 in subjects with cystic fibrosis homozygous for F508del: results from a phase II study (FLAMINGO)
Kors Van der Ent (Utrecht, Netherlands)
14:36 - 14:44

Safety, pharmacokinetics and pharmacodynamics of the CFTR corrector FDL169
Alexander Horsley (Manchester, United Kingdom)
14:44 - 14:52

Safety, tolerability and pharmacokinetics of the CFTR potentiator FDL176
Claudia Ordonez (Charlestown, United States)
14:52 - 15:00

ePoster Session
14:00 - 15:00
ePoster Corners

ePS2, Genotype/phenotype correlations: CFTR and other disease modifiers

ePoster Corner B
Harriet Corvol (Paris, France)
Michael Wilschanski (Jerusalem, Israel)

Introduction
14:00 - 14:04

Association of polymorphic variants of GSTT1, GSTM1, GSTP1 and GCLC genes with the severity of the cystic fibrosis clinical manifestations
Olga G. Novoselova (Moscow, Russian Federation)
14:04 - 14:11

Influence of phase 1 xenobiotics metabolism genes on the efficiency of antibacterial therapy in patients from the Russian Federation and the Republic of Belarus
Elena Kondratyeva (Moscow, Russian Federation)
14:11 - 14:18

The influence of polymorphism of metalloproteinase genes on the formation of cystic fibrosis-liver cirrhosis and cystic fibrosis pneumofibrosis
Anastasia Goryainova (Moscow, Russian Federation)
14:18 - 14:25

Combination of liver cirrhosis with mild mutation p.E92K
Yulia Gorinova (Moscow, Russian Federation)
14:25 - 14:32

Genotype/phenotype correlation in cystic fibrosis children with mutation 3272-16T>A
Vera Shadrina (Perm, Russian Federation)
14:32 - 14:39

Phenotypic manifestations of rare missense mutations
Yulia Gorinova (Moscow, Russian Federation)
14:39 - 14:46
**Scientific Programme**

**Intestinal current and nasal potential difference index cases: diagnostic features of subjects with CFTR-related disorder**
Burkhard Tümmler (Hanover, Germany)

**Clinical characteristics of patients with one known cystic fibrosis-related mutation despite extended genotype testing**
Christopher Brockelsby (Liverpool, United Kingdom)

---

**ePoster Session**
14:00 - 15:00
ePoster Corners

**ePS1, Airway epithelial cells to study cystic fibrosis pathogenesis and find new therapies**
ePoster Corner A

Tanja Gonska (Toronto, Canada)
Nicoletta Pedemonte (Genoa, Italy)

**Chronic rhinosinusitis: reduced Ca2+-mediated Cl- secretion observed in vitro is confirmed by nasal potential difference measurements in patients**
Johanna Salomon (Heidelberg, Germany)

**Biochemical and biophysical properties of bronchial mucus from preschool children with cystic fibrosis**
Kathryn Ramsey (Bern, Switzerland)

**Human respiratory epithelial cells prevent Aspergillus fumigatus germination**
Nicolas Richard (Paris, France)

**Use of ex vivo paediatric primary nasal epithelial cultures to investigate TMEM16A as a potential therapeutic target in children with cystic fibrosis**
Iram Haq (Newcastle upon Tyne, United Kingdom)

**CAT-5571 restores autophagy, a fundamental defect in cystic fibrosis, and is a potential new treatment for people with cystic fibrosis**
Feng Liu (Cambridge, United States)

**Overproduction of IL-6 and TNF through IRE1α is CFTR genotype dependent**
Samuel Lara Reyna (Leeds, United Kingdom)

---

**Poster Viewing 1**

**Differential gene expression by Pseudomonas aeruginosa in response to bacteriophage infection**
Rossa Brugha (London, United Kingdom)
Whole genome variant analysis and transposon sequencing provides insight into aztreonam resistance in cystic fibrosis Pseudomonas aeruginosa airway infection
Kathryn Mclean (Seattle, United States)

In vivo confirmation of Pseudomonas aeruginosa biofilms as independent pharmacological compartments - implication for antibiotic treatment
Lars Christophersen (Copenhagen, Denmark)

Intraclonal competitive fitness of serial Pseudomonas aeruginosa isolates from cystic fibrosis lungs
Burkhard Tümmler (Hanover, Germany)

Monitoring of chronic Pseudomonas aeruginosa lung infection in cystic fibrosis patients
Lusine R. Avetisyan (Moscow, Russian Federation)

Carbapenem-resistant shared Pseudomonas aeruginosa strains with oprD mutations in cystic fibrosis
Laura Sherrard (Belfast, United Kingdom)

Multi-drug resistant Pseudomonas aeruginosa strains in cystic fibrosis and FEV1 decline
Daniela Dolce (Florence, Italy)

A 6-year longitudinal population study of Pseudomonas aeruginosa from initial colonisation in the Montpellier CF centre, France
Chloé Dupont (Montpellier, France)

Antimicrobial resistance of cystic fibrosis Pseudomonas aeruginosa: a longitudinal study from initial colonisation in cystic fibrosis patients with special focus on temocillin
Chloé Dupont (Montpellier, France)

Azithromycin resistance develops fast in P. aeruginosa but has no negative impact on lung function development in cystic fibrosis patients with chronic infection
Oana Ciofu (Copenhagen, Denmark)

Strain and antibiogram changes, over time, in paediatric cystic fibrosis patients colonised with Pseudomonas aeruginosa
Dervla Kenna (London, United Kingdom)

Detection of Pseudomonas aeruginosa in exhaled breath of cystic fibrosis patients
Christof Majoor (Amsterdam, Netherlands)

‘Dr Dog’ will see you now? First steps in assessing the utility of trained sniffer dogs to detect Pseudomonas aeruginosa (Pa) airway infection in non-expectorating cystic fibrosis patients
Jane Davies (London, United Kingdom)

Phenotyping and molecular typing of Achromobacter spp. strains in children and adults with cystic fibrosis in the Russian Federation
Marina Yu Chernukha (Moscow, Russian Federation)

Chronic infection with Achromobacter xylosoxidans in paediatric and adult cystic fibrosis patients in two centres in Argentina
Ezequiel Baran (La Plata, Argentina)
Contribution of *Stenotrophomonas maltophilia* in colonisation of the respiratory tract in cystic fibrosis patients
Hanna Dmeńska (Warsaw, Poland)

Late impact of early chronic methicillin-sensitive *Staphylococcus aureus* (MSSA) infection in children with cystic fibrosis
FRANCOIS GALODE (BORDEAUX, France)

Use of extended-quantitative culture does not predict an imminent pulmonary exacerbation
Laura Sherrard (Belfast, United Kingdom)

Human reservoirs of pathogens colonising the airways of cystic fibrosis patients
Rebeca Passarelli Mantovani (Verona, Italy)

A cross-sectional analysis of cough swabs from non-expectorating cystic fibrosis patients and healthy control participants
Gisli Einarsson (Belfast, United Kingdom)

Brain abscess caused by *Escherichia coli* in an adult with cystic fibrosis
Ezequiel Baran (La Plata, Argentina)

Macrophage activation syndrome due to *Nocardia* in a cystic fibrosis patient
Tugba Sismanlar Eyuboglu (Ankara, Turkey)

Epidemiology of nontuberculous mycobacteria at an adult cystic fibrosis centre
Tegan Greener (Liverpool, United Kingdom)

The epidemiology of nontuberculous mycobacteria in a paediatric cystic fibrosis centre
Ana Kotnik Pirš (Ljubljana, Slovenia)

The use of aprepitant as part of an effective anti-emetic regimen to facilitate successful completion of intensive intravenous *Mycobacterium abscessus* therapy
Bogumila Belkarty (London, United Kingdom)

Surveillance for ototoxicity in children with *Mycobacterium abscessus* lung disease in a UK cystic fibrosis centre
Sally Palser (Nottingham, United Kingdom)

Extended SABC incubation for *Exophiala* species in cystic fibrosis sputa
Evelyn Ioi (Greater Manchester, United Kingdom)

Clinical and immunological results of the Berlin Aspergillus Species Study (BASS)
Carsten Schwarz (Berlin, Germany)

Berlin Aspergillus Species Study (BASS) - pet ownership is a risk factor for ABPA in patients with cystic fibrosis
Svenja Kaufmann (Berlin, Germany)

*Aspergillus fumigatus* degrades the respiratory mucins MUC5B and MUC5AC
Alexander Horsley (Manchester, United Kingdom)
Diverse clinical characteristics of Aspergillus growth in patients with cystic fibrosis  
Nagehan Emiralioglu (Ankara, Turkey)

Presence of Candida dubliniensis in the cystic fibrosis respiratory tract was associated with a significant decline in lung function - results from a 16-year retrospective study  
Mahasin Al Shakirchi (Stockholm, Sweden)

Changes in airways bacterial community with cystic fibrosis patients’ age and lung function decline  
Olga Voronina (Moscow, Russian Federation)

Longitudinal analysis of the lower airway microbiota in cystic fibrosis patients during clinical stability compared to those experiencing episodes of pulmonary exacerbation  
Gisli Einarsson (Belfast, United Kingdom)

Predicting the functional intestinal profiles in cystic fibrosis  
Chee Y. Ooi (Randwick, Australia)

Taxonomic and functional microbial signatures of cystic fibrosis lung disease  
Annamaria Bevivino (Rome, Italy)

Use of qPCR to analyse changes in total bacterial and Pseudomonas aeruginosa load in cystic fibrosis patients when clinically stable and during exacerbations  
Clodagh McGettigan (Belfast, United Kingdom)

Information value of non-invasive specimens for airway bacteriome research  
Olga Voronina (Moscow, Russian Federation)

Anti-fungal therapeutic drug monitoring in adults with cystic fibrosis  
Kavita Dave (London, United Kingdom)

What should you do after stopping flucloxacillin in children?  
Iolo Doull (Cardiff, United Kingdom)

Discontinuation of prophylactic flucloxacillin in young adults with cystic fibrosis  
William Flight (Oxford, United Kingdom)

Activity of liposomal loaded azithromycin against cystic fibrosis clinical respiratory isolates  
Rachel Mairs (Belfast, United Kingdom)

The impact of non-antimicrobial therapies on bacterial pathogens commonly found in cystic fibrosis airways  
Zara Kirkwood (Belfast, United Kingdom)

Potential taste disturbance is largely mild and diminishes with continuing treatment in cystic fibrosis patients treated with levofloxacin inhaled solution  
Stuart Elborn (London, United Kingdom)
**Scientific Programme**

- **Effect of inhaled ceftazidime on early Burkholderia contaminans lung infection in children with cystic fibrosis**
  Virginia D'Alessandro (La Plata, Argentina)

- **Inhaled dry powder colistin (Colobreathe®) in regular cystic fibrosis therapy in patients from a centre in Sofia**
  Guergana Petrova (Sofia, Bulgaria)

- **Inhaled anti-pseudomonal therapy in patients with cystic fibrosis**
  Andreas Hector (Tübingen, Germany)

- **Bacteriophage treatment of multidrug-resistant Pseudomonas aeruginosa pneumonia in a cystic fibrosis patient**
  Carrie-Lynn Furr (San Diego, United States)

- **Synergistic activity of colistin in combination with N-acetylcysteine against colistin-resistant Acinetobacter baumannii grown in biofilms**
  Francesco Sergio (Milan, Italy)

- **Characterisation of colistimethate sodium delivery from different nebuliser systems**
  Dearbhla Hull (Chichester, United Kingdom)

- **Monitoring amikacin pharmacokinetics for personal dose optimisation**
  Regina Leshem (Jerusalem, Israel)

- **Synergistic activities of an efflux pump inhibitor and antibiotics against Pseudomonas aeruginosa isolates from cystic fibrosis patients**
  Douweh Gbian (Sudbury/Ontario, Canada)

- **In vitro susceptibility to ceftolozane/tazobactam in Pseudomonas aeruginosa isolates at an adult cystic fibrosis centre**
  Gemma Edwards (Wythenshawe, United Kingdom)

- **Midline administration of aminoglycoside antibiotics is safe in adult cystic fibrosis patients**
  Nichola MacDuff (Wolverhampton, United Kingdom)

- **A service evaluation addressing: are intravenous antibiotics administered at home as clinically effective as intravenous antibiotics administered in hospital in cystic fibrosis patients in the adult cystic fibrosis centre in Aberdeen?**
  Sandra Steele (Aberdeen, United Kingdom)

- **Oral versus intravenous treatment of bronchopulmonary exacerbations in cystic fibrosis**
  Carsten Schwarz (Berlin, Germany)

- **Design, enrollment, and feasibility of the STOP-2 randomised study of intravenous antibiotic treatment duration in cystic fibrosis pulmonary exacerbations**
  Patrick Flume (Charleston, United States)

- **Intravenous fosfomycin for treatment of pulmonary exacerbations in adult cystic fibrosis patients with chronic infection by Pseudomonas aeruginosa and other Gram-negative bacteria**
  Barbara Messore (Orbassano (TO), Italy)
Influenza vaccination in children with cystic fibrosis using live attenuated influenza vaccine: experience from five seasons in an Austrian children CF centre
Sabine Renner (Vienna, Austria)

Role of an exacerbation checklist score in the definition, assessment and outcome of cystic fibrosis pulmonary exacerbations
Charlotte Addy (Belfast, United Kingdom)

The “rare-exacerbator” status in adult cystic fibrosis patients
Andrea Gramegna (Milan, Italy)

Pneumococcal antibody levels in cystic fibrosis patients attending the All Wales Adult CF Centre
Annette Smith (Vale of Glamorgan, United Kingdom)

Staphylococcal infection impact on the outcome of lung disease in cystic fibrosis
Elena Roxana Smâdeanu (Bucharest, Romania)

Clinical data of patients with nontuberculous mycobacteria in a paediatric cystic fibrosis centre in Slovenia
Marina Praprotnik (Ljubljana, Slovenia)

First Aspergillus fumigatus seroconversion was associated with more severe disease in cystic fibrosis children
Hortense Petat (ROUEN, France)

Clinical effects of omalizumab in adult cystic fibrosis patients with allergic severe asthma associated with allergic bronchopulmonary aspergillosis
Barbara Messore (Orbassano (TO), Italy)

Prevalence, clinical characteristics and outcomes of patients referred to an adult cystic fibrosis centre from pulmonary clinics
Martina Contarini (Milan, Italy)

Clinical outcomes associated with infected totally implantable venous access devices
Paul Griffiths (Liverpool, United Kingdom)

Outcome of adults with cystic fibrosis admitted to the medical intensive care unit
Berrin Er (Ankara, Turkey)

Heart rate as predictor of cystic fibrosis severity
Filipa Ferro (Lisboa, Portugal)

Cell-free DNA and DNase I activity in cystic fibrosis
Elena Kondratyeva (Moscow, Russian Federation)

In vitro effects of rhDNase on sputum in patients with cystic fibrosis using rheology
Jérémy Patarin (Saint Martin d’Hères, France)

In vivo effects of rhDNase and saline solution on sputum in patients with cystic fibrosis using rheology
Jérémy Patarin (Saint Martin d’Hères, France)
Testing sputum in patients with cystic fibrosis using rheology: the ageing effect
Jérémy Patarin (Saint Martin d'Hères, France)

Exhaled nitric oxide is a possible biomarker for predicting complications in cystic fibrosis patients
Andreas Renner (Wien, Austria)

Plasma sphingomyelin and ceramide levels of cystic fibrosis patients
Berrin Er (Ankara, Turkey)

How does the basement membrane change in patients with cystic fibrosis?
Vaclav Koucky (Prague, Czech Republic)

Risk factors for early lung function decline in young children with cystic fibrosis
Alejandro Teper (Buenos Aires, Argentina)

Lung function and its association with nutritional and functional status in patients with cystic fibrosis living in the Moscow region
Yulia Gorinova (Moscow, Russian Federation)

Time course of pulmonary hyperinflation in adult cystic fibrosis: clinical and lung function correlates
Camille Audousset (Lille, France)

Differences in molecular diffusivity do not explain discrepancy in Lung Clearance Index measured by nitrogen and SF6 washout
Carl Whitfield (Manchester, United Kingdom)

An 8-week open-label interventional multi-centre study to evaluate the Lung Clearance Index as endpoint for clinical trials in cystic fibrosis patients ≥ 6 years of age, chronically infected with Pseudomonas aeruginosa
Matthias Welsner (Essen, Germany)

Lung Clearance Index measurement in children with primary ciliary dyskinesia and cystic fibrosis; Hacettepe University experience
Nagehan Emiralioglu (Ankara, Turkey)

Short-term effect of physiotherapy on Lung Clearance Index among patients with cystic fibrosis
Elpis Hatziagorou (Thessaloniki, Greece)

Change in the Lung Clearance Index (LCI) with microbiological status in children with cystic fibrosis - pilot study in a new paediatric cystic fibrosis centre in Poland
Katarzyna Walicka-Serzysko (Warsaw, Poland)

Lung Clearance Index and vitamin D
Ioana Mihaiea Ciucu (Timisoara, Romania)

Bicarbonate and oxygen saturation predict the need for fitness to fly test in patients with cystic fibrosis
Giulia Spoletini (Leeds, United Kingdom)
Scientific Programme

An audit of the use of computerised tomography scans for chest imaging in a large UK paediatric cystic fibrosis clinic
Clare Halfhide (Liverpool, United Kingdom)

Bronchoscopy and airway clearance techniques in cystic fibrosis
Helga Elidottir (Lund, Sweden)

The role of the specialist cystic fibrosis pharmacist in embedding adherence monitoring of inhaled therapies into clinical care
C McKeown (Belfast, United Kingdom)

Lung transplantation in paediatric patients with cystic fibrosis: a single centre experience
Elena Spinelli (Verona, Italy)

Immunosuppression in cystic fibrosis: how worried should we be?
Elizabeth Clarke (Manchester, United Kingdom)

Persistence of peribronchial tertiary lymphoid structures in cystic fibrosis patients treated with rituximab
Lucile Regard (Paris, France)

A qualitative account of the first UK cystic fibrosis trust physiotherapy fellowship
Rachael Bass (Newcastle Upon Tyne, United Kingdom)

Re-audit of the UK ACPCF Physiotherapy National Standards of Care
Lisa Morrison (Glasgow, United Kingdom)

Teaching adult patients CF physiotherapy: a French experience
Claudine Lejosne-Artige (RENNES, France)

Improving physiotherapy practice with educational videos
Kevin Cobb (Vienna, Austria)

CF@home: feasibility and utility of ear lobe blood gas sampling in the patient’s home
Kate Channon (London, United Kingdom)

Retrospective clinical data on IPV therapy in children with cystic fibrosis
Peter Anderson (Glasgow, United Kingdom)

Sampling methods used in non-expectorating children ≤3 years of age with cystic fibrosis in a London cystic fibrosis centre
Jasmine Miller (London, United Kingdom)

Vestibular dysfunction and aminoglycoside exposure in cystic fibrosis
Sara Keane (Dublin, Ireland)

The minimal clinically important difference for survival in patients with cystic fibrosis
Wytze Doeleman (Utrecht, Netherlands)

The impact of exacerbations on muscle force, exercise capacity and quality of life in adults with cystic fibrosis
Filip Pyl (Ghent, Belgium)
Isokinetic evaluation of maximum quadriceps strength in 37 cystic fibrosis adult patients  
Sophie Ramel (Roscoff, France)

A multidimensional analysis of exercise capacity amongst adults with cystic fibrosis  
Ronan Buckley (Dublin, Ireland)

Exercise: love it, hate it, or somewhere in-between? Identifying exercise self-efficacy and the impact of personal training in children and young people with cystic fibrosis  
Helen Douglas (London, United Kingdom)

Development of Hacka Health 4 CF: a physiotherapy technology solution to support young people with cystic fibrosis  
Sarah Rand (London, United Kingdom)

Impact of the optimal duration of intensive rehabilitation training on patients with moderate to severe cystic fibrosis  
Hugues Gauchez (Marcq en Baroeul, France)

Virtual exercise class for cystic fibrosis inpatients - a novel way to encourage exercise participation  
Fiona Haynes (Nottingham, United Kingdom)

Service evaluation of a community exercise programme for people with cystic fibrosis  
Fiona Haynes (Nottingham, United Kingdom)

Expansion of a leading cystic fibrosis exercise programme: collaborative partnership between a UK health and fitness provider and five UK cystic fibrosis centres  
Ashleigh Ahlquist (London, United Kingdom)

Reasons for non-compliance with cardiopulmonary exercise testing in cystic fibrosis  
Jayne Trott (Exeter, United Kingdom)

Feasibility of following up the benefits of a rehabilitation program by telehomecare in children with cystic fibrosis  
Caroline Bruneaux (Bordeaux, France)

Cardiopulmonary exercise test and lumacaftor/ivacaftor in cystic fibrosis  
Lue K. D. Philipsen (Copenhagen, Denmark)

Service evaluation of an out-patient exercise programme for cystic fibrosis patients  
Fiona Haynes (Nottingham, United Kingdom)

An interim service evaluation of App-based individual exercise programmes which are remotely monitored and progressed by a clinical physiologist specialising in exercise from a large Adult Cystic Fibrosis Centre  
Ian Waller (Manchester, United Kingdom)

Feasibility of using online video calling to engage patients in the management of cystic fibrosis  
Owen Tomlinson (Exeter, United Kingdom)
Mobile phone step-counter data does not correlate with objective measures of exercise capacity
Paul Griffiths (Liverpool, United Kingdom)

Role of osteopathic manipulative treatment in cystic fibrosis: a pilot study
Valentina Fainardi (Parma, Italy)

Demographic and clinical data of Croatian patients with cystic fibrosis - first results from the adult care cystic fibrosis centre
Ivona Markelić (Zagreb, Croatia)

Clinical and genetic characteristics of patients in a Lisbon Cystic Fibrosis Centre - importance in the face of CFTR targeted therapies
Teresa Rodrigues (Lisboa, Portugal)

Characteristics of cystic fibrosis in regions of the Russian Federation
Natalia Ilienkova (Krasnoyarsk, Russian Federation)

Searching for CFTR mutations in healthy children and cystic fibrosis paediatric patients from Russian populations
Rena Zinchenko (Moscow, Russian Federation)

Characteristics of the frequency of homozygotes F508del according to the register of patients with cystic fibrosis of the Russian Federation in 2016
Elena Kondratyeva (Moscow, Russian Federation)

Searching for CFTR mutations in CF patients from North Ossetia-Alania Republic, Russian Federation
Tatyana A. Vasilyeva (Moscow, Russian Federation)

Data from the Cystic Fibrosis Patient Registry of the Russian Federation (comparison 2011 and 2016)
Nataliya Kashirskaya (Moscow, Russian Federation)

The characteristics of chronic respiratory infection by Pseudomonas aeruginosa in the Cystic Fibrosis Patient Registry in the Russian Federation in 2016
Victoria Sherman (Moscow, Russian Federation)

The health status of children with cystic fibrosis in Macedonia according to the Cystic Fibrosis Registry in 2015
Tatjana Jakovska-Maretti (Skopje, Macedonia, the Republic of)

Analysis of demographic characteristics and cumulative survival of cystic fibrosis population in the Republic of Macedonia
Stojka Fushtik (Skopje, Macedonia, the Republic of)

Incidence of cystic fibrosis in Paraguay
Marta Ascurra (Asunción, Paraguay)

Early clinical determinants of respiratory outcome in patients with cystic fibrosis
Julian Forton (Cardiff, United Kingdom)
Scientific Programme

Pulmonary exacerbations and risk of death amongst patients with cystic fibrosis (CF) with homozygous and heterozygous F508del mutations in Ireland
Mariam Hassan (Boston, United States)

Pulmonary function patterns and their association with genotype and phenotype in adult cystic fibrosis patients
Dimitri Stylemans (Brussel, Belgium)

Cystic Fibrosis-Related Diabetes before lung transplantation impact on survival and long-term renal function
Quitterie Reynaud (Lyon, France)

Malignant diseases in patients treated at the CF Centre Innsbruck
Dorothea Appelt (Innsbruck, Austria)

The health status of mothers with CF in Bulgaria
Guergana Petrova (Sofia, Bulgaria)

Human papilloma virus vaccination among female patients at the Auvergne-Rhône Alpes paediatric cystic fibrosis centres
Isabelle Durieu (Lyon, France)

I'm with the band
Georgina Aldous (Liverpool, United Kingdom)

Canadian Pharmacists’ and Patients’ Perspectives on Cystic Fibrosis Pharmacist Roles in an Outpatient Multidisciplinary Care Team
Nadia McTaggart (Victoria, Canada)

Quality improvement in Registry data entry: experience from a UK centre
Nicola Pinnock (Liverpool, United Kingdom)

Cystic fibrosis sub-speciality training: will supply meet demand?
Paul Griffiths (Liverpool, United Kingdom)

ECFS Tomorrow Session
14:00 - 15:00

Managing challenging behaviour in young people with CF during in-patient stays
Kath MacDonald (Edinburgh, United Kingdom)
Ann Raman (Ghent, Belgium)

Workshop
15:00 - 16:30

WS01, Exciting news from CFTR modulator clinical trials
Clinical Trials / New Therapies
Silke van Koningsbruggen-Rietschel (Cologne, Germany)
Lieven Dupont (Leuven, Belgium)
Scientific Programme

A phase 3, 2-part, single-arm study of ivacaftor treatment in patients < 2 years with a CFTR gating mutation: results from the ARRIVAL study in patients 1 to 2 years
Margaret Rosenfeld (Seattle, United States)

Phase 2 initial results evaluating PTI-428, a novel CFTR amplifier, in patients with cystic fibrosis
Patrick Flume (Charleston, United States)

Translational read-through of CFTR nonsense mutations and inducement of cystic fibrosis transmembrane conductance regulator (CFTR) function by ELX-02 treatment
Neal Sharpe (Waltham, United States)

GLPG2222 in subjects with cystic fibrosis and the F508del/Class III mutation on stable treatment with ivacaftor: results from a phase II study (ALBATROSS)
Scott Bell (Brisbane, Australia)

A 2-part, phase 3, single-arm study to evaluate the safety and pharmacokinetics (PK) of lumacaftor/ivacaftor (LUM/IVA) combination therapy in patients (pts) aged 2 to 5 years with cystic fibrosis homozygous for the F508del-CFTR mutation
Gregory Sawicki (Boston, United States)

Preliminary safety and efficacy of triple combination CFTR modulator regimens in cystic fibrosis
Jane Davies (London, United Kingdom)

Workshop
15:00 - 16:30
Hall 1

WS02, NTM: what's in the clinic, new diagnostics and treatments
Microbiology / Antibiotics
Andres Floto (Cambridge, United Kingdom)
Marianne Skov (Copenhagen, Denmark)

Epidemiology of nontuberculous mycobacteria infection in children and young people with cystic fibrosis: analysis of United Kingdom Cystic Fibrosis Trust Registry data
Malcolm Brodlie (Newcastle upon Tyne, United Kingdom)

Prevalence of nontuberculous mycobacteria in potable water
Rebecca Stockwell (Brisbane, Australia)

Antibody testing for Mycobacterium avium complex infection in cystic fibrosis patients
Cecilie Ravnholt (Copenhagen, Denmark)

Glycopeptidolipids of the Mycobacterium abscessus cell wall are immunodominant antigens and represent potential targets for a diagnostic assay
Carsten Schwarz (Berlin, Germany)
Scientific Programme

Treatment success for eradication of pulmonary nontuberculous mycobacterial infections in a paediatric cystic fibrosis cohort  16:00 - 16:15
Dominic Hughes (London, United Kingdom)

Improving antibiotic activity against Mycobacterium abscessus isolates by inducing bacterial disaggregation and oxygen availability 16:15 - 16:30
Mette Kolpen (Copenhagen, Denmark)

Interactive Poster Discussions 15:00 - 16:30  Amphitheatre

IPD1, Mental health in cystic fibrosis: assessment and care

Mandy Bryon (London, United Kingdom)
Urszula Borawska - Kowalczyk (Warsaw, Poland)

Mental health screening in cystic fibrosis centres across Europe
Janice Abbott (Preston, United Kingdom)

Assessing strengths and difficulties as part of annual mental health screening
Alistair Duff (Leeds, United Kingdom)

Depression among parents of children with cystic fibrosis and its socioeconomic determinants
Renata Zubrzycka (Lublin, Poland)

Implementing the International Committee on Mental Health in Cystic Fibrosis (ICMH) guidelines: practical complexities of managing mental health
Marieke Verkleij (Amsterdam, Netherlands)

Instituting mental health screening in a Canadian paediatric cystic fibrosis clinic
Anna Gravelle (Vancouver, BC, Canada)

Outcome of mental screening among Swedish adolescents with cystic fibrosis
Carolina Laine (Stockholm, Sweden)

Cystic fibrosis: does the deterioration of pulmonary function affect the level of anxiety and depression?
Ivana Lalic (Zagreb, Croatia)

Treating unresolved grief in parents of children with cystic fibrosis
André Schultz (Perth, Australia)

Workshop 15:00 - 16:30  Hall 2

WS03, Towards optimal nutritional status

Anne Munck (Paris, France)
Jochen G. Mainz (Jena, Germany)
Scientific Programme

**Growth, body composition and lung function in pre-pubertal children with cystic fibrosis diagnosed via newborn screening, and comparison with a historical unscreened cohort**
Elizabeth Owen (London, United Kingdom)

**The effect of an intensive residential rehabilitation program with adjusted nutritional care on body composition in adult patients with cystic fibrosis**
Stefanie Dereeper (De Haan, Belgium)

**Childhood nutritional status is a major factor determining lung function in adults with cystic fibrosis**
Moshe Ashkenazi (Ramat Gan, Israel)

**Comparing abdominal symptoms in cystic fibrosis patients and healthy controls with a novel multimodal questionnaire (CF-Abd Score)**
Anke Jaudszus (Jena, Germany)

**The human intestinal proteome in children with cystic fibrosis**
Michael Coffey (Sydney, Australia)

**A first approach for an evidence-based method to adjust PERT: in vivo validation of the in vitro model**
Joaquim Calvo-Lerma (Valencia, Spain)

**Workshop**
15:00 - 16:30

**WS04, New ways to modulate inflammation in cystic fibrosis**

**Immunology / Pulmonology / Inflammation**

Pierre-Régis Burgel (Paris, France)
Thomas Scambler (Leeds, United Kingdom)

**Targeting IL-17-producing T-cells attenuates the severity of Pseudomonas aeruginosa lung infection**
Cristina Cigana (Milan, Italy)

**Azithromycin treatment of Pseudomonas aeruginosa infection potentiates IgY pulmonary protection in vivo**
Kim Thomsen (Copenhagen, Denmark)

**Sodium influx modulates innate immune inflammation and metabolism in cystic fibrosis**
Thomas Scambler (Leeds, United Kingdom)

**Investigation of the CFTR-TGFβ1 interaction regarding inflammatory effects in wildtype and CFTR-mutated human bronchial epithelial cells**
Jan Christoph Thomassen (Cologne, Germany)

**The effect of cysteamine and epigallocatechin gallate on hyperresponsive inflammation in cystic fibrosis**
Jonathan Holbrook (Leeds, United Kingdom)
Balancing the immune response in cystic fibrosis: using zebrafish models of inflammation to uncover new therapeutic approaches
Audrey Bernut (Sheffield, United Kingdom)

16:15 - 16:30

Workshop
15:00 - 16:30
Annex B

WS05, Which factors influence outcomes from newborn to adulthood?

Epidemiology / Registry
Siobhan Carr (London, United Kingdom)
Christian Benden (Zurich, Switzerland)

Impact of newborn screening on outcomes and social inequalities in cystic fibrosis: a UK Registry-based study
Daniela K. Schlüter (Lancaster, United Kingdom)

Impact of tobacco smoke exposure on pulmonary function in paediatric cystic fibrosis patients
Gabriela Oates (Birmingham, United States)

New Pseudomonas chronicity score: evaluating association with clinical outcomes and comparing to Leeds criteria
Margaret Rosenfeld (Seattle, United States)

Clinical effectiveness results from the first interim analysis of the VOCAL study: an observational study of ivacaftor in patients with cystic fibrosis and selected non-G551D gating mutations
Carlo Castellani (Verona, Italy)

Pregnancy outcome in women with cystic fibrosis and poor pulmonary function
Quitterie Reynaud (Lyon, France)

Mortality outcomes related to multi-drug resistant organisms in cystic fibrosis lung transplant recipients: an International Society of Heart and Lung Transplantation (ISHLT) thoracic transplant Registry study
Christian Benden (Zurich, Switzerland)

15:00 - 15:15
15:15 - 15:30
15:30 - 15:45
15:45 - 16:00
16:00 - 16:15
16:15 - 16:30

Symposium
17:00 - 18:30
Blue Hall

SS01, Standards of care: moving ahead

Pavel Drevinek (Prague, Czech Republic)
Martin Stern (Tübingen, Germany)

The implementation of best practice procedures around Europe
Stefano Aliberti (Milan, Italy)

Evaluation of the quality of CF care from the patients’ perspective
Vincent Gulmans (Baarn, Netherlands)

17:00 - 17:22
17:22 - 17:44
Scientific Programme

Benchmarking in Scandinavia
 Tacjana Pressler (Copenhagen, Denmark) 17:44 - 18:06

Organisation of CF care: the Serbian experience
 Predrag Minic (Belgrade, Serbia) 18:06 - 18:30

WS06, Assessing lung structure and function in cystic fibrosis

Harm Tiddens (Rotterdam, Netherlands)
Elpis Hatziagorou (Thessaloniki, Greece)

Monitoring cystic fibrosis lung disease in school-age children with computer tomography
 Marcus Svedberg (Gothenburg, Sweden) 17:00 - 17:15

Interest of sequential expiratory chest computed tomography in monitoring lung disease of children with cystic fibrosis
 Marie Mittaine (Toulouse, France) 17:15 - 17:30

A novel approach based on low field nuclear magnetic resonance to monitor lung functionality and inflammation in cystic fibrosis patients
 Michela Abrami (Trieste, Italy) 17:30 - 17:45

Lung ultrasound score and the relation with lung clearance index
 Ioana Mihaiela Ciucu (Timisoara, Romania) 17:45 - 18:00

A methodology to evaluate trapped gas using an open circuit multiple breath nitrogen washout (MBWN2) device
 Christopher Short (London, United Kingdom) 18:00 - 18:15

Effect of lumacaftor/ivacaftor on lung clearance index and exercise capacity among patients aged over 12 years with cystic fibrosis homozygous for F508del-CFTR
 Elpis Hatziagorou (Thessaloniki, Greece) 18:15 - 18:30

WS07, Challenging cases in 21st century cystic fibrosis

Kath MacDonald (Edinburgh, United Kingdom)
Pavla Hodkova (Prague, Czech Republic)

Moving into the 21st century with dietetic care - changing patient perceptions
 Emma Farrell (London, United Kingdom) 17:00 - 17:15
Scientific Programme

Emergent biopsychosocial challenges of novel drug development and clinical trial participation: stopping an experimental modulator drug in order to conceive
Rebecca Dobra (London, United Kingdom)

The lonely patient: "I survived, now what?"
Edwina Landau (Petah Tikva, Israel)

Bereavement and loss in a 14 year-old girl and the impact on cystic fibrosis
Trudy Havermans (Leuven, Belgium)

Raising the medication intake in a kindergarten setting - a case report
Angela Gabele (Zürich, Switzerland)

At a loss: the failings of multi-agency working?
Holly Clisby (London, United Kingdom)

Workshop
17:00 - 18:30

WS08, Exercise: the fitter the better!

Physiotherapy

Helge Hebestreit (Würzburg, Germany)
Marta Kerstan (Biberstein, Switzerland)

The utility of oxygen uptake efficiency as a marker of aerobic fitness in children with cystic fibrosis
Owen Tomlinson (Exeter, United Kingdom)

Acute effects of combined exercise and oscillatory positive expiratory pressure therapy on sputum properties and lung diffusing capacity in cystic fibrosis: a randomised, controlled, crossover trial
Thomas Radtke (Zurich, Switzerland)

Combined ramp and supramaximal cardiopulmonary exercise testing for individuals with cystic fibrosis
Zoe Saynor (Portsmouth, United Kingdom)

Feasibility of an incremental step test to assess aerobic fitness in adult cystic fibrosis
Zelda Beverley (London, United Kingdom)

Balance, flexibility and agility - additional aspects of physical fitness and trainability in children and adolescents with cystic fibrosis
Wolfgang Gruber (Essen, Germany)

Effects of home-based adapted physical activity in patients with cystic fibrosis: an interventional study
Boubou Camara (Grenoble, France)
WS09, Pharmacology and genetic tools for cystic fibrosis basic research correction

Basic Science
Patrick T. Harrison (Cork, Ireland)
Rob Tarran (Chapel Hill, United States)

- **Development of a CFTR mRNA therapy capable of treating lung disease in all patients with cystic fibrosis**
  Ann Barbier (Cambridge, United States)
  17:00 - 17:18

- **Developing scarless and permanent CFTR correction using CRISPRs**
  Jim Hu (Toronto, Canada)
  17:18 - 17:36

- **Development of effective method for F508del mutation correction using CRISPR/Cas9**
  Alexander Vlavrov (Moscow, Russian Federation)
  17:36 - 17:54

- **Targeting αENaC with an epithelial RNAi trigger delivery platform for the treatment of cystic fibrosis**
  Erik Bush (Madison, United States)
  17:54 - 18:12

- **The ENaC regulatory peptide SPX-101 is stable in CF sputum and functional regardless of CFTR mutation**
  David Scott (Durham, United States)
  18:12 - 18:30

WS10, Screening around the globe: from foetus to newborn

Genetics / Screening / Diagnosis
Milan Jr Macek (Prague, Czech Republic)
Emmanuelle Girodon (Paris, France)

- **Isolated non-visualisation of foetal gallbladder should be considered for the prenatal diagnosis of cystic fibrosis**
  Emmanuelle Girodon (Paris, France)
  17:00 - 17:15

- **International approaches for delivery of positive newborn bloodspot screening results for cystic fibrosis**
  Jane Chudleigh (London, United Kingdom)
  17:15 - 17:30

- **Cystic fibrosis newborn screening in Austria: after 20 years changing the algorithm from IRT/IRT to IRT/PAP/IRT**
  Sabine Renner (Vienna, Austria)
  17:30 - 17:45

- **Evaluating cystic fibrosis newborn screening diagnostic performance in the United States**
  Susanna A. McColley (Chicago, United States)
  17:45 - 18:00

- **The Irish Comparative Outcome Study (ICOS): clinical outcomes at 3 years following introduction of newborn cystic fibrosis screening**
  Patricia Fitzpatrick (Dublin, Ireland)
  18:00 - 18:15
Scientific Programme

Seven years of nationwide cystic fibrosis newborn screening in the Czech Republic: analysis of the IRT/DNA/IRT scheme outcomes
Milan Jr Macek (Prague, Czech Republic)
Symposium
08:30 - 10:00

S13, Infection control and transmissibility in cystic fibrosis - what is clear and what is not?

Upon completion of this session participants should have:
- An overview of how CF infection control guidelines evolved and their importance in preventing cross-infection with CF pathogens
- Detailed knowledge of recent outbreaks of CF pathogens in Eastern Europe and lessons learnt from these outbreaks
- A focused understanding of disinfecting protocols as part of optimal infection prevention strategies in CF healthcare environments
- A detailed understanding of strategies to prevent aerosol transmission of CF pathogens

Tavs Qvist (Copenhagen, Denmark)
Michael Tunney (Belfast, United Kingdom)

How the CF infection control guidelines evolved
Andrew Jones (Manchester, United Kingdom) 08:30 - 08:52

CF pathogen outbreaks in Eastern Europe
Marina Yu Chernukha (Moscow, Russian Federation) 08:52 - 09:14

Disinfection protocols to prevent transmission of bugs
Dieter Worlitzsch (Halle, Germany) 09:14 - 09:36

Strategies to prevent aerosol transmission in cystic fibrosis
Timothy Kidd (Brisbane, Australia) 09:36 - 10:00

Symposium
08:30 - 10:00

S14, Early airway disease - how aggressive should we be?

Upon completion of this session participants should be able to:
- Understand the pathogenesis of early airway disease
- Learn about how to diagnose and monitor early airway disease
- Appreciate the complexities of treating early airway disease

Kris De Boeck (Leuven, Belgium)
Eitan Kerem (Jerusalem, Israel)

Pathogenesis of early airway disease
Isabelle Sermet (Paris, France) 08:30 - 08:52

Identification and monitoring of early disease - how aggressive should we be?
Eitan Kerem (Jerusalem, Israel) 08:52 - 09:14

Treatment of early disease - how aggressive should we be?
Marijke Proesmans (Leuven, Belgium) 09:14 - 09:36
**How will we manage CFTR modulators?**

Philippe Reix (Lyon, France)

09:36 - 10:00

---

**Symposium**

08:30 - 10:00

**Amphitheatre**

**S15, Which bacteria require eradication in cystic fibrosis?**

Upon completion of this session participants should be able to:

- Learn about the potential environmental sources of NTM infection
- Understand how whole genome sequencing can inform the epidemiology of NTM infection
- Get an overview of current treatment outcomes and discuss how we might approach NTM management in the future

Mieke Boon (Leuven, Belgium)
Predrag Minic (Belgrade, Serbia)

08:30 - 08:52

**Pseudomonas aeruginosa**

Giovanni Taccetti (Florence, Italy)

08:52 - 09:14

**Stenotrophomonas maltophilia**

Christine Roenne Hansen (Lund, Sweden)

09:14 - 09:36

**Achromobacter**

Lieven Dupont (Leuven, Belgium)

09:36 - 10:00

**MRSA**

Marianne S. Muhlebach (Chapel Hill, United States)

---

**Symposium**

08:30 - 10:00

**Hall 2**

**S16, Whose patient is it anyway? Managing differences of opinion between MD teams and parents/patients**

Upon completion of this session participants should be able to:

- Consider ethical, moral and medical decision-making processes within the CF Team
- Discuss the CF teams’ relationships with parents and people with CF and the ways in which decision-making is negotiated in these relationships
- Be informed about developmental stages of young people with CF and consider how this might influence behaviours, treatment choices and decision-making

Janice Abbott (Preston, United Kingdom)
Jacqueline Noordhoek (Baarn, Netherlands)

08:30 - 08:52

**Point: when is too little care too much harm?**

Mandy Bryon (London, United Kingdom)

08:52 - 09:14

**Counterpoint: too little care or too little collaboration?**

Gregory Sawicki (Boston, United States)

09:14 - 09:36

**Audience discussion (Pro/Con)**


09:36 - 10:00

**Psychological support for young people who push the boundaries**

Katrien Van Gompel (Antwerp, Belgium)
Scientific Programme

Symposium
08:30 - 10:00

Annex A

S17, Body composition - aiming for the lean?
Upon completion of this session participants should be able to:
- Learn the importance of a good nutritional assessment in the management of cystic fibrosis
- Understand the multidisciplinarity of rebuilding lean body mass in cystic fibrosis

Sarah Collins (London, United Kingdom)
Dimitri Declercq (Ghent, Belgium)

We should routinely measure body composition in all patients - PRO
Susannah King (Melbourne, Australia)
08:30 - 08:48

We should routinely measure body composition in all patients - CON
Chris Smith (Brighton, United Kingdom)
08:48 - 09:06

Discussion
09:06 - 09:16

The influence of CFTR-modulators on nutritional status
Tanja Gonska (Toronto, Canada)
09:16 - 09:38

Lean body mass from the perspective of a physiologist
Craig Williams (Exeter, United Kingdom)
09:38 - 10:00

Annex B

S18, Novel analysis of CF Registry data
Upon completion of this session participants should be able to:
- Identify innovative uses of CF Registries
- Appraise the wealth of information contained in CF Registries
- Discover additional uses of epidemiological data sets

Ruth Keogh (London, United Kingdom)
Alexander Elbert (Bethesda, United States)

Understanding health inequalities with Registry data
Daniela K. Schlüter (Lancaster, United Kingdom)
08:30 - 08:52

Can analysis of data using machine learning guide clinical decisions?
Thomas Daniels (Southampton, United Kingdom)
08:52 - 09:14

Longitudinal, multi-country comparisons of CF Registry data
Christopher Goss (Seattle, United States)
09:14 - 09:36

Longitudinal FEV₁% predicted using ECFSPR data
Elpis Hatziagorou (Thessaloniki, Greece)
09:36 - 10:00
S19, Can we predict response to new therapies?

Upon completion of this session participants should be able to:
- Describe the different methods of epithelial cell culture
- Explain the characteristics of epithelial organoids
- Compare pros and cons of the different models available for in vitro CFTR drug testing

Tim Lee (Leeds, United Kingdom)
Jane Davies (London, United Kingdom)

- Nasal epithelial cells for CFTR drug testing
  Iwona Pranke (Paris, France)

- Stem cells as sources for CF disease modeling and drug discovery
  Massimo Conese (Foggia, Italy)

- Intestinal vs. airway organoids
  Jeffrey Beekman (Utrecht, Netherlands)

- New methodologies to implement clinical trials
  Kris De Boeck (Leuven, Belgium)

S20, Upper airway disease management

Upon completion of this session participants should be able to:
- Understand the role of sinonasal infection in the acquisition and persistence of P. aeruginosa airway infection in CF
- Understand the medical and surgical treatment options for upper airway disease
- Learn about optimal management of the upper airway before and after lung transplantation

Jochen G. Mainz (Jena, Germany)
Tacjana Pressler (Copenhagen, Denmark)

- The sinonasal role in acquisition and persistence of P. aeruginosa in CF airways
  Jochen G. Mainz (Jena, Germany)

- Conservative treatment
  Assen Koitschev (Stuttgart, Germany)

- Surgical treatment
  Ron Eliashar (Jerusalem, Israel)

- Management before and after transplantation
  Letizia Corinna Morlacchi (Milan, Italy)
Scientific Programme

Symposium
10:30 - 12:00  Amphithéatre

S21, CF diagnosis - doing more good than harm

Upon completion of this session participants should be able to:
- Remember the weaknesses and strengths of different CF NBS protocols
- Interpret how different algorithms will influence the number of infants detected with CFSPID
- Assess the value of a CFSPID registry

Jürg Barben (St. Gallen, Switzerland)
Elke De Wachter (Brussels, Belgium)

Weaknesses and strengths of PAP in NBS  10:30 - 10:52
Olaf Sommerburg (Heidelberg, Germany)

Weaknesses and strengths of DNA analysis in NBS  10:52 - 11:14
Emmanuelle Girodon (Paris, France)

The challenges and opportunities of having a CFSPID registry  11:14 - 11:36
Anne Munck (Paris, France)

Ambiguous clinical presentations - benefits and pitfalls of making a CF diagnosis  11:36 - 12:00
Ana Kotnik Pirš (Ljubljana, Slovenia)

Symposium
10:30 - 12:00  Hall 2

S22, Ensuring standards of psychosocial care all across Europe: training, education, mentoring

Upon completion of this session participants should be able to:
- Recognise the East-West divide in application of The European Standards of CF Care
- Identify the differences in CF management across cultures
- Construct creative ways how teams can work together to promote best practice for social and mental wellbeing of people with CF and their families

Pavla Hodkova (Prague, Czech Republic)
Anna Elderton (Oxford, United Kingdom)

What does good mental health care look like - Western perspective  10:30 - 10:52
Marieke Verkleij (Amsterdam, Netherlands)

What does good mental health care look like - Eastern perspective  10:52 - 11:14
Urszula Borawska - Kowalczyk (Warsaw, Poland)

Training, education, mentoring  11:14 - 11:36
Alistair Duff (Leeds, United Kingdom)

Social care expectations: East vs. West  11:36 - 12:00
Suja Chandran (London, United Kingdom)
Scientific Programme

 Symposium
 10:30 - 12:00

 S23, Interactive case studies
 Eitan Kerem (Jerusalem, Israel)
 Giulia Spoletini (Leeds, United Kingdom)

 10:30 - 10:40
 Complicated Cystic Fibrosis-Related Diabetes: non-adherence or a new medication?
 Claire Edmondson (London, United Kingdom)

 10:40 - 10:50
 Renal involvement in an adult patient with cystic fibrosis
 Emma Luke (London, United Kingdom)

 10:50 - 11:00
 A CF infant with complicated neonatal course - a diagnostic and management dilemma
 Laura Fawcett (Sydney, Australia)

 11:00 - 11:10
 Is this cystic fibrosis?
 Marija Dimitrovska-Ivanova (Stip, Macedonia, the Republic of)

 Symposium
 10:30 - 12:00

 S24, The epithelial channelome and alternative strategies for rescue

 This session focuses on alternative strategies for CF therapy. Speakers will focus on the potential therapeutic relevance of targeting novel CFTR regulators to improve F508del maturation and trafficking to the plasma membrane, as well as modulating non-CFTR channels and transporters to help restore chloride and bicarbonate secretion to improve airways hydration and pH

 Michael Gray (Newcastle upon Tyne, United Kingdom)
 Nicoletta Pedemonte (Genoa, Italy)

 10:30 - 10:52
 Novel CFTR regulators and their potential roles as drug targets
 Nicoletta Pedemonte (Genoa, Italy)

 10:52 - 11:14
 Manipulating acid-base transporters as a therapeutic approach for cystic fibrosis lung disease
 Vinciane Saint-Criq (Newcastle upon Tyne, United Kingdom)

 11:14 - 11:36
 CFTR and SLC26A9 interaction: prospects for therapy
 Marcus Mall (Berlin, Germany)

 11:36 - 12:00
 Rescue of the chloride permeability defect in CF by alternate ion channels
 Martin Gosling (Falmer, United Kingdom)

 Satellite Symposium
 12:30 - 14:00

 Satellite Symposium

 Please find the detailed program of the Satellite Symposia here.
Satellite Symposium
12:30 - 13:30

Satellite Symposium
Please find the detailed program of the Satellite Symposia here.

Meet the Experts
12:45 - 13:45
ePoster Corners

Meet the Experts 4: Pregnancy
ePoster Corner A
Jennifer Taylor-Cousar (Denver, United States)
Malena Cohen-Cymerknoh (Jerusalem, Israel)

Meet the Experts 5: How to do a paper review?
ePoster Corner B
Scott Bell (Brisbane, Australia)
Patrick Flume (Charleston, United States)
Carlo Castellani (Verona, Italy)

Meet the Experts 6: Aging
ePoster Corner C
Nicholas Simmonds (London, United Kingdom)
Pierre-Régis Burgel (Paris, France)

ECFS Tomorrow Session
12:45 - 13:45

Body Composition - Practical considerations and techniques
Elizabeth Owen (London, United Kingdom)
ePoster Session
14:00 - 15:00
ePoster Corners

ePS5, Emerging insights from Registries
ePoster Corner B
Margaret Rosenfeld (Seattle, United States)
Andreas Jung (Zurich, Switzerland)

Introduction
14:00 - 14:04

Personalised time-updated predictions of short-term and long-term survival in cystic fibrosis using UK Registry data
Ruth Keogh (London, United Kingdom)

Estimation of survival of cystic fibrosis patients in France by two different methods
Virginie Scotet (Brest, France)

Stenotrophomonas maltophilia and Achromobacter xylosoxidans in cystic fibrosis: epidemiology and lung disease progression
Linda Bouhlel (Nice, France)

The role of Pseudomonas infection in early growth trajectories in cystic fibrosis
Amy Macdougall (London, United Kingdom)

Epidemiology and phenotype of cystic fibrosis with residual function mutations in Italy
Donatello Salvatore (Potenza, Italy)

Demography and clinical outcomes in cystic fibrosis lung transplant recipients in Belgium
Muriel Thomas (Brussels, Belgium)

Evaluating the impact of 2006 clinical practice guidelines for nutrition in children with cystic fibrosis in Australia
Rasa Ruseckaite (Melbourne, Australia)

Quality improvement in CF: what can we learn from each other? A statistical analysis of UK Registry data and consultations with clinicians and patients
Stephanie Macneill (Bristol, United Kingdom)

ePoster Session
14:00 - 15:00
ePoster Corners

ePS4, Microbiome interactions and updates on key pathogens
ePoster Corner A
Tavs Qvist (Copenhagen, Denmark)
Debby Bogaert (Edinburgh, United Kingdom)

Introduction
14:00 - 14:04
Scientific Programme

**Microbial community composition in explanted cystic fibrosis lungs**
Christopher D. Spence (Belfast, United Kingdom)

14:04 - 14:11

**Porphyromonas, a candidate biomarker for detection of Pseudomonas aeruginosa pulmonary infection in cystic fibrosis**
Geneviève Héry-Arnaud (Brest, France)

14:11 - 14:18

**Clinical implications of the cystic fibrosis airway microbial metagenome**
Burkhard Tümmler (Hanover, Germany)

14:18 - 14:25

**Characterising the intestinal virome in cystic fibrosis**
Michael Coffey (Sydney, Australia)

14:25 - 14:32

**Determining the relationship between absolute quantification and relative abundance as determined by next generation sequencing**
Clodagh McGettigan (Belfast, United Kingdom)

14:32 - 14:39

**Mutual antagonism: a complex coexistence of Aspergillus fumigatus and Pseudomonas aeruginosa in the cystic fibrosis airway**
Emma Reece (Dublin, Ireland)

14:39 - 14:46

**Interactions of Pseudomonas aeruginosa with other bacterial species in an artificial sputum model**
Laura Wright (Liverpool, United Kingdom)

14:46 - 14:53

**NTM infection in paediatrics: how do we approach these emerging pathogens?**
Gemma Saint (Liverpool, United Kingdom)

14:53 - 15:00

---

ePoster Session
14:00 - 15:00
ePoster Corners

**ePS6, Mixing up physiotherapy**

**ePoster Corner C**

Jamie Wood (Perth, Australia)
Peter Suter (Basel, Switzerland)

**Introduction**

14:00 - 14:04

**Shaking it up: a look at our centre experience with high frequency chest wall oscillation (HFCWO)**
Carwyn Bridges (Cardiff, United Kingdom)

14:04 - 14:11

**Investigating the experience of adults with cystic fibrosis using long-term domiciliary non-invasive ventilation**
Jocelyn Choyce (Birmingham, United Kingdom)

14:11 - 14:18

**Hypertonic saline use in our paediatric cystic fibrosis centre: using the UK cystic fibrosis Registry to reflect on practice**
Rachel O'Connor (London, United Kingdom)

14:18 - 14:25

**Correlation between six-minutes-walk-test and cystic fibrosis disease severity**
Elad Ben-Meir (Ramat Gan, Israel)

14:25 - 14:32
A-STEP: a maximal, incremental, externally paced step test for adults with cystic fibrosis - safety and feasibility testing  
Lisa Wilson (Melbourne, Australia)

Influence of a supervised exercise program (CFmobil) on motor ability in adult cystic fibrosis patients  
Matthias Welsner (Essen, Germany)

Effects of physical retraining on the body composition of adult patients with cystic fibrosis  
Anne Prevotat (Lille, France)

The effects of Orkambi therapy on muscle strength, function, body composition and quality of life in cystic fibrosis patients homozygous for CFTR-F508del  
Clare M. Reilly (Dublin, Ireland)

ePoster Session  
14:00 - 15:00

ePoster Viewing 2

Two decades of experience in prenatal diagnosis and carrier screening of cystic fibrosis in Serbia  
Danijela Radivojevic (Belgrade, Serbia)

Importance of population-specific first step CFTR screening in Serbian cystic fibrosis patients  
Dragica Radojkovic (Belgrade, Serbia)

Newborn screening for cystic fibrosis in Serbia: a pilot study  
Sanja Grkovic (Belgrade, Serbia)

Results of a 3-year screening in Konya province and the frequency in our region  
Sevgi Pekcan (Konya, Turkey)

Two years of newborn screening for cystic fibrosis in Turkey: Cukurova experience  
Derya Ufuk Altintas (adana, Turkey)

Advanced search for CFTR gene mutations in Russian cystic fibrosis patients  
Nika Petrova (Moscow, Russian Federation)

Current situation of genotyping of cystic fibrosis patients in the Russian Federation  
Nataliya Kashirskaya (Moscow, Russian Federation)

CFTR gene mutation spectrum peculiarities in Russian cystic fibrosis patients  
Tagui Adyan (Moscow, Russian Federation)

Fast detection of CFTR mutations in dried blood spots of newborns in the Khanty-Mansi region (Russia)  
Maxim Donnikov (Surgut, Russian Federation)
Scientific Programme

The efficiency of cystic fibrosis newborn screening for homozygous F508del patients
Victoria Sherman (Moscow, Russian Federation)

Cystic fibrosis newborn screening: a Belgian experience
Hedwige BOBOLI (LIEGE, Belgium)

Comparison of 2 strategies of newborn screening for cystic fibrosis: IRT/IRT vs. IRT/PAP
Alejandro Teper (Buenos Aires, Argentina)

Selection of a genetic test for neonatal screening of cystic fibrosis in the mixed population of Luxembourg
Marizela Kulisic (Dudelange, Luxembourg)

Genetic study of patients with cystic fibrosis in a mixed population of northeastern Brazil
Edna Lúcia Souza (savior, Brazil)

First molecular report of the ΔF508 mutation in patients with cystic fibrosis in Paraguay
Marta Ascurra (Asunción, Paraguay)

Late diagnosis of cystic fibrosis - the possible hazard for the family
Guergana Petrova (Sofia, Bulgaria)

A bioethical framework for the evaluation of newborn bloodspot screening for cystic fibrosis
Rachael Armstrong (Wirral, United Kingdom)

A comparative study between clinically diagnosed and screen-detected children with cystic fibrosis: parents’ economic costs prior to diagnosis
Patricia Fitzpatrick (Dublin, Ireland)

Advanced molecular dynamics simulations for understanding the functions and dysfunctions of the CFTR channel
Isabelle Callebaut (Paris, France)

Development of a 3D full thickness cystic fibrosis model on chip
Claudia Mazio (Napoli, Italy)

A phase 3, open-label study of tezacaftor/ivacaftor (TEZ/IVA) therapy: interim analysis of pooled safety, and efficacy in patients homozygous for F508del-CFTR
Patrick Flume (Charleston, United States)

Safety, tolerability and pharmacokinetics of CFTR corrector FDL169
Claudia Ordonez (Charlestown, United States)

Impact of lumacaftor/ivacaftor (Orkambi®) on proton pump inhibitor use in adult cystic fibrosis patients
Anna Connolly (Dublin, Ireland)

Effects of food and moderate hepatic impairment on the pharmacokinetics (PK) of tezacaftor/ivacaftor (TEZ/IVA)
Lakshmi Viswanathan (Boston, United States)
Lumacaftor/ivacaftor in real life for Phe508del homozygous adolescents with severe and normal lung function
Cyrielle Collet (Bordeaux, France)

Real-world Orkambi Cohort Cork study (ROCKS) - a prospective three months' analysis addressing the impact of CFTR modulation in patients with cystic fibrosis homozygous for ΔF508del CFTR
Parniya Arooj (Cork, Ireland)

Alteration of lipid epoxy metabolites due to CFTR modulation with lumacaftor/ivacaftor in df508del homozygous patients
Jobst Roehmel (Berlin, Germany)

Riociguat for the treatment of adult Phe508del homozygous cystic fibrosis: efficacy data from the Phase II Rio-CF study
Jennifer Taylor-Cousar (Denver, United States)

Upper airway symptoms in cystic fibrosis - is dornase alfa administered by pulsating aerosol a treatment option?
Fiona Haynes (Nottingham, United Kingdom)

Modelled deposition of colistimethate sodium aerosol in the lungs of patients with cystic fibrosis using two different mesh nebulisers
Dearbhla Hull (Chichester, United Kingdom)

Home registrations of inhalations with tobramycin inhalation powder - the relation between inspiratory flow and cough
Jennifer Meerburg (Rotterdam, Netherlands)

The analgesic effect of salmon calcitonin on acute rib fractures in cystic fibrosis
Annabelle Lee (London, United Kingdom)

A twinned remote program of supervised adapted physical activity (APA) for French & Irish people living with cystic fibrosis improves self-reported physical activity level and physical fitness
Elodie Desplanche (Dooradoyle, Co. Limerick, Ireland)

Are patients with cystic fibrosis in clinical trials sensitive to the placebo effect? A metanalysis
Julie Coton (Lyon, France)

Patient-friendly glucose tolerance test for patients with cystic fibrosis
Sigrid Amstelveen-Bokkerink (Groesbeek, Netherlands)

Usefulness of extended oral glucose tolerance test in patients with cystic fibrosis older than 16 years
Ezequiel Baran (La Plata, Argentina)

Striking the balance between cystic fibrosis and Cystic Fibrosis-Related Diabetes (CFRD) - a meta-ethnography
Sarah Collins (London, United Kingdom)

The Cystic Fibrosis-Related Diabetes self-management triad - it is about more than just numbers
Sarah Collins (London, United Kingdom)
The relationship of Cystic Fibrosis-Related Diabetes (CFRD) and abnormal lipid profiles in adults with cystic fibrosis
Steven Caskey (Belfast, United Kingdom)

Cystic Fibrosis-Related Diabetes, chronic lung infection and low nutritional status increase the risk of cystic fibrosis-related bone disease
Ana Kotnik Pirš (Ljubljana, Slovenia)

How can continuous glucose monitoring system impact on therapy of Cystic Fibrosis-Related Diabetes (CFRD) in youth
Sladjana Todorovic (Belgrade, Serbia)

Hyperglycemia may predispose to lower respiratory tract infection in young children with cystic fibrosis
Bernadette Prentice (Randwick, Australia)

The use of dipeptidyl peptidase-4 (DPP-4) inhibitors in cystic fibrosis
Emma Farrell (London, United Kingdom)

Understanding the mechanisms for the increased prevalence of gastro-oesophageal reflux in cystic fibrosis
Robert Lord (Manchester, United Kingdom)

The relationship between gastro-oesophageal reflux and cystic fibrosis lung disease
Robert Lord (Manchester, United Kingdom)

Improved clinical outcomes following Nissen fundoplication in cystic fibrosis patients: experience in a large adult cystic fibrosis centre
Kavita Sivabalah (Liverpool, United Kingdom)

Differences in clinical outcomes of paediatric cystic fibrosis patients with and without meconium ileus
Chee Y. Ooi (Randwick, Australia)

How to give pancreatic enzyme in cystic fibrosis children with gastrostomy and unable to swallow opotherapy by mouth?
Claire Collard (Liege, Belgium)

Colorectal cancer in adults with cystic fibrosis: our experience at the All Wales Adult Cystic Fibrosis Centre
Anna Sayers (Penarth, United Kingdom)

Lactase activity in small intestinal biopsies in children with cystic fibrosis
Elena Roslavtseva (Moscow, Russian Federation)

Faecal calprotectin in cystic fibrosis patients during respiratory exacerbation
Ze’ev Schnapp (Haifa, Israel)

Cystic fibrosis-related fatty liver disease is associated with Pseudomonas aeruginosa infection and reduced lung function
Charlotte Wrey (London, United Kingdom)

Features of clinical status in children with cystic fibrosis complicated by liver cirrhosis and portal hypertension in the Russian Federation
Olga Malomuzh (Moscow, Russian Federation)
Scientific Programme

Is APRI index evaluation useful in diagnosing hepatic changes in the course of cystic fibrosis?
Sabina Wiecek (Katowice, Poland)

Sodium status in children with cystic fibrosis: evaluation of fractional sodium excretion in a paediatric cohort
Marijke Proesmans (Leuven, Belgium)

A retrospective audit of sodium chloride supplementation in infants with cystic fibrosis
Katherine Stead (Birmingham, United Kingdom)

Salt supplementation in CF: too much or too low?
Shrabani Chakraborty (London, United Kingdom)

Vitamin E supplementation with fat soluble formulation in children with cystic fibrosis: two years follow-up
Juliana Roda (Coimbra, Portugal)

An analysis of vitamin K supplementation during the first five years of life in paediatric cystic fibrosis patients managed at Addenbrooke’s hospital, Cambridge
Sophie Kelly (Cambridge, United Kingdom)

Identification of subclinical deficit of vitamin K in cystic fibrosis by Micro-Raman spectroscopy
Juliana Roda (Coimbra, Portugal)

Factors influencing total serum cholesterol in cystic fibrosis children and adolescents - preliminary data
Monika Mielus (Warsaw, Poland)

The feasibility and applicability of the Healthy Eating Index for Australian Adults (HEIFA-2013) score for dietary assessment in adults with cystic fibrosis
Susannah King (Melbourne, Australia)

Children in Bulgaria with mainly gastrointestinal onset of cystic fibrosis have worse nutritional results, despite being diagnosed earlier
Guergana Petrova (Sofia, Bulgaria)

Evolution of body composition measured by BIA after the start of tube feeding
Stephanie Van Biervliet (Ghent, Belgium)

Bio-electrical impedance analysis and relationship with pulmonary function in cystic fibrosis patients
Francis Hollander (Utrecht, Netherlands)

Body composition measured by BIA of tube-fed cystic fibrosis patients compared to age-sex matched cystic fibrosis controls
Stephanie Van Biervliet (Ghent, Belgium)

Body composition changes in the first year of treatment with lumacaftor-ivacaftor in adults with severe cystic fibrosis lung disease
Susannah King (Melbourne, Australia)
Gender differences in lipid profiles in adults with cystic fibrosis
Steven Caskey (Belfast, United Kingdom)

To be or not to be (happy): fat is the question?
Jamie Duckers (Cardiff, United Kingdom)

Cost analysis of the treatment of exacerbations in children with cystic fibrosis depending on nutritional status
Elena Zhekaite (Moscow, Russian Federation)

Evaluating the effectiveness of dietetic interventions: developing a dietetic outcome measures tool for Cystic Fibrosis
Laurie Cave (Leeds, United Kingdom)

Relationship between nutritional status and pulmonary function in adults with cystic fibrosis: cross-sectional and longitudinal analyses
Francis Hollander (Utrecht, Netherlands)

Breastfeeding and higher SES lead to better outcomes in children with cystic fibrosis
Albert Faro (Bethesda, United States)

FODMAP intake and gastrointestinal symptoms in cystic fibrosis - is there a relationship?
Audrey Tierney (Melbourne, Australia)

Evaluation of the effect of dietary therapy after PEG insertion on anthropometric and respiratory indices in patients with cystic fibrosis
Paulina Jazgarska (Rabka - Zdrój, Poland)

Nutritional status and eating behaviour in a cystic fibrosis population
Tiago Martins (Lisbon, Portugal)

Food diary analysis and growth: do they correlate? A pilot study
Chris Smith (Brighton, United Kingdom)

Features of somatic growth of children with cystic fibrosis in different age periods in the Russian Federation
Anna Voronkova (Moscow, Russian Federation)

How well do children with cystic fibrosis grow in a cystic fibrosis centre in Sweden when no newborn screening exists, but an old tradition with physical activity as the baseline treatment?
Mikael Nilsson (Lund, Sweden)

Malnutrition in adolescent and adult patients with cystic fibrosis
Lidija Spirevska (Skopje, Macedonia, the Republic of)

An increase in weight and fat mass observed following five months of ivacaftor treatment plateaus at 24 months in adults with G551D-related cystic fibrosis
Audrey Tierney (Melbourne, Australia)

Influences on vitamin D in cystic fibrosis patients
Mihaela Dediu (Timisoara, Romania)

Putting some backbone into managing low vitamin D
Dawn Lau (Vale of Glamorgan, United Kingdom)
Scientific Programme

The influence of genetic factors and phenotype on the development of vitamin D deficiency in children with cystic fibrosis
Elena Zhekaite (Moscow, Russian Federation)

Vitamin D deficiency in patients with cystic fibrosis: are we adhering to monitoring recommendations?
Susan Jacob (Seattle, United States)

Monitoring bone health in the All Wales Adult CF Centre (AWACFC)
Chloe Wilson (Penarth, United Kingdom)

Inflammation and bone turnover markers in cystic fibrosis
Inger H. Mathiesen (Copenhagen, Denmark)

Prevalence of low BMD, osteoporosis and DEXA scan attendance in the North Ireland Adult CF Centre: the relationship to current guidelines
Veronica Lynch (Belfast, United Kingdom)

Cardiovascular risk in an adult cystic fibrosis population: a 10-year retrospective analysis
Nick Greig (Leeds, United Kingdom)

The heart in cystic fibrosis - three cases from Bulgaria
Guergana Petrova (Sofia, Bulgaria)

Vascular function in adults with cystic fibrosis
James Shelley (Liverpool, United Kingdom)

Age at menarche in girls with cystic fibrosis
Marie Mittaine (Toulouse, France)

Fertility in women with cystic fibrosis: a French survey
Marie Mittaine (Toulouse, France)

Cystic fibrosis with pancreatic insufficiency is associated with a high incidence of subfertility in women
Michal Shteinberg (Haifa, Israel)

Partnering with patients and parents of children with cystic fibrosis in the French care quality improvement program
Dominique Bertrand (BOBIGNY, France)

What do parents want from annual review?
Jacqui Cowlard (London, United Kingdom)

Pharmacist consultation introduced to annual review of adults with cystic fibrosis
Michaela Smith (Christchurch, New Zealand)

Home intravenous antibiotics: behind closed doors
Annette Smith (Vale of Glamorgan, United Kingdom)

Home delivery influences medicines possession ratio in adult cystic fibrosis
Michael Dooney (Manchester, United Kingdom)
Scientific Programme

Totally implantable venous access device tip position and risk of venous complications in cystic fibrosis
Amie Hart (Oxford, United Kingdom)

Aseptic centralised versus home extemporaneous preparation for cystic fibrosis outpatients' parenteral antibiotic therapy: a survey on nurses' satisfaction
Stephan Garcia (Saint-Genis Laval, France)

Support to self-esteem building among children aged 6 to 10 years old: ”My little notebook that makes me feel better”
Pilar Leger (Nantes, France)

My day... my week: a new therapeutic educational tool in Liège Cystic Fibrosis Centre
Marc-Antoine Wuidart (Liège, Belgium)

Using poetry to teach students and others about cystic fibrosis
Kath MacDonald (Edinburgh, United Kingdom)

Genetic counselling: do our patients know what we think they know?
Chloe Wilson (Penarth, United Kingdom)

Round table meetings for parents of cystic fibrosis babies diagnosed through newborn screening
Annemarie Tiesinga (Groningen, Netherlands)

An attempt to reduce the psychological consequences of isolation during paediatric hospitalisations in a cystic fibrosis centre
Urszula Borawska-Kowalczyk (Warsaw, Poland)

Adolescents with cystic fibrosis and the risk of acquiring and transmitting respiratory germs
Suzy Gonsseaume (Paris, France)

How the Socratic questioning method can be used by the cystic fibrosis multidisciplinary team to develop a deeper understanding of patients' difficulties and aid decision-making
Isobelle Jasmin Biggin (Bristol, United Kingdom)

Developing awareness of psychosocial issues for the cystic fibrosis multidisciplinary team - using the psychologist differently
Bethan Phillips (Cardiff, United Kingdom)

Transplant eligibility perceptions amongst a cystic fibrosis multidisciplinary team
Christopher Brockelsby (Liverpool, United Kingdom)

The cost of caring: nursing experiences of palliation within cystic fibrosis care
Clare Sumner (Liverpool, United Kingdom)

Life coaching among young adults with cystic fibrosis - a qualitative study
Karin Bæk Knudsen (Copenhagen, Denmark)
Scientific Programme

Views of healthcare professionals on patients having access to their secondary care electronic healthcare record: results of a cross-sectional questionnaire
Akhil Sawant (Leeds, United Kingdom)

The use of an online learning module (LearnPro NHSTM) to educate and assess staff working in cystic fibrosis care
Aileen Mallinson (Edinburgh, United Kingdom)

Pioneering virtual reality technology for transition at the All Wales Adult CF Centre
Lorraine Speight (Vale of Glamorgan, United Kingdom)

Association between adherence to azithromycin and dornase alpha and lung function decline in adult cystic fibrosis patients: a two-year analysis
Natalia Popowicz (Perth, Australia)

Why won't children keep exercising? Barriers and facilitators to exercise and physical activity maintenance - a qualitative study
Helen Douglas (London, United Kingdom)

Evaluation of youth work support for teenagers and young adults with cystic fibrosis
Adnan Mehmood (Leeds, United Kingdom)

Fertility and fatherhood in men with cystic fibrosis
Alan Anderson (Newcastle upon Tyne, United Kingdom)

Quality of life in cystic fibrosis: influence of some psychosocial factors on disease severity
Tatjana Zorcec (Skopje, Macedonia, the Republic of)

Quality of life and influencing factors in children and young adults with cystic fibrosis
Bulent Karadag (Istanbul, Turkey)

Anxiety and depression in adult patients with cystic fibrosis in Croatia: results from an adult cystic fibrosis centre
Ivana Lalic (Zagreb, Croatia)

Factors related to anxiety and depression in adult cystic fibrosis patients
Nazli Zeynep Uslu (Istanbul, Turkey)

Comparison of depression, burnout, caregiver burden and parent-child attitudes in cystic fibrosis and primary ciliary dyskinesia in children’s mothers
Tugba Sismanlar Eyuboglu (Ankara, Turkey)

Outcome measures as suggested by the cystic fibrosis community
Alan Smyth (Nottingham, United Kingdom)

How does the current clinical trials landscape reflect the James Lind Alliance top ten research priorities for cystic fibrosis?
Alan Smyth (Nottingham, United Kingdom)

When doctors go to patients: a patient-based evaluation of the CFQ
Rosa Coucke (Paris, France)
Scientific Programme

**Living with cystic fibrosis - are we all forgetting something?**
Lorraine Speight (Vale of Glamorgan, United Kingdom)

**Impact of cystic fibrosis on healthy siblings: a systematic review**
Jane Chudleigh (London, United Kingdom)

**Symposium**
15:00 - 16:30

**SS02, Special Symposium 2 - The Early Cystic Fibrosis Years**
Kevin Southern (Liverpool, United Kingdom)
Kris De Boeck (Leuven, Belgium)

- **Navigating the psychological pitfalls of the early CF years** 15:00 - 15:22
  Trudy Havermans (Leuven, Belgium)

- **Achieving nutritional excellence in partnership with parents** 15:22 - 15:44
  Ruth Watling (United Kingdom)

- **How to sample the airway, how to use inhaled medicines in preschool children** 15:44 - 16:06
  Felix Ratjen (Toronto, Canada)

- **Introducing new therapies to small children safely and promptly** 16:06 - 16:30
  Isabelle Sermet (Paris, France)

**Workshop**
15:00 - 16:30

**WS11, Novel treatments for bacterial biofilms and bad bugs**
Microbiology / Antibiotics

Barbara Kahl (Münster, Germany)
Laura Sherrard (Belfast, United Kingdom)

- **Alginate oligomers as novel therapies to treat life-threatening pseudomonal multidrug resistant bacterial infections** 15:00 - 15:15
  Juliette Oakley (Cardiff, United Kingdom)

- **Hyperbaric oxygen treatment enhances the effect of tobramycin against biofilm-growing Pseudomonas aeruginosa** 15:15 - 15:30
  Signe Agnete Møller (Copenhagen Ø, Denmark)

- **GVF27, HVA36 and IMY47: new human derived host defence peptides for alternative cystic fibrosis therapeutic intervention strategies** 15:30 - 15:45
  Elio Pizzo (Naples, Italy)

- **The repurposed multiple sclerosis drug, glatiramer acetate, is an antibiotic resistance breaker in Pseudomonas aeruginosa strains from cystic fibrosis patients** 15:45 - 16:00
  Ronan A. Murphy (London, United Kingdom)
41st European Cystic Fibrosis Conference,
07 - 10 June 2017,
Belgrade, Serbia

Scientific Programme

**Post-antibiotic effect of hypothiocyanite, lactoferrin and ALX-009 on clinical strains isolated from cystic fibrosis patients**
Victor Juarez Perez (Lyon, France)

**The effect of 2-thiopyridine derivative 11026103, a novel antibacterial compound, on Burkholderia cenocepacia: transcriptomic response and resistance mechanisms**
Jaroslav Nunvar (Prague, Czech Republic)

---

Workshop
15:00 - 16:30
Amphitheatre

**WS12, How cold electronics can help to provide warmth of care**

*Nursing / Psychosocial Issues*

Daniel Peckham (Leeds, United Kingdom)
Joaquim Calvo-Lerma (Valencia, Spain)

**Pioneering virtual reality technology for distraction therapy at the All Wales Adult CF Centre**
Anna Sayers (Penarth, United Kingdom)

**Supporting cystic fibrosis care at University Hospital Limerick (UHL) through eHealth**
Niamh Murphy (Limerick, Ireland)

**Telehealth monitoring for home intravenous antibiotics in cystic fibrosis**
Laura Blanch (Newcastle upon Tyne, United Kingdom)

**Are goal-focused, motivational text messages effective at improving patients' belief in their own ability (self-efficacy) to complete inhaled therapies in adults with cystic fibrosis**
Kate Channon (London, United Kingdom)

**A smartphone application for reporting symptoms in adults with cystic fibrosis: a randomised controlled trial**
Jamie Wood (Perth, Australia)

**MyCyFAPP project: validation of the PEDsQL GI symptom scale to evaluate gastro-intestinal symptoms in children with cystic fibrosis**
Mieke Boon (Leuven, Belgium)

---

Workshop
15:00 - 16:30
Hall 2

**WS13, Assessing the patient: it's not all about the chest**

*Physiotherapy*

Andrew Jones (Manchester, United Kingdom)
Fiona Cathcart (London, United Kingdom)
Scientific Programme

15:00 - 15:15
Thoracic ultrasound as a useful tool to assess lung atelectasic areas and set the best physiotherapy
Matteo Giuliari (Rovereto, Italy)

15:15 - 15:30
Musculoskeletal symptoms in adults with cystic fibrosis
Elizabeth Clarke (Manchester, United Kingdom)

15:30 - 15:45
The impact of pain on physiotherapy care in cystic fibrosis
Anna Meschi (Verona, Italy)

15:45 - 16:00
Experiences of non-invasive ventilation in a large adult cystic fibrosis centre and nationally
Zelda Beverley (London, United Kingdom)

16:00 - 16:15
Assessment of the prevalence and severity of urinary and ano-rectal functional disorders and their impact on quality of life and sexuality in adult with cystic fibrosis in the French North-West Cystic Fibrosis Network
Sophie Ramel (Roscoff, France)

16:15 - 16:30
Urinary incontinence in adult patients with cystic fibrosis: prevalence, impact on daily life and relationship with chronic cough
Ernesto Crisafulli (Parma, Italy)

Workshop
15:00 - 16:30
Annex A

WS14, Clinical trials on lung inflammation, lung clearance and exercise
Clinical Trials / New Therapies

15:00 - 15:15
ROSCO-CF, a safety and efficacy clinical trial of (R)-roscovitine in cystic fibrosis patients
Laurent Meijer (Roscoff, France)

15:15 - 15:30
Phase 3 randomised controlled study of the efficacy and safety of inhaled mannitol in adults with cystic fibrosis
Elena Amelina (Moscow, Russian Federation)

15:30 - 15:45
Randomised, double-blind, controlled pilot study on safety and efficacy of hypertonic saline as preventive inhalation therapy in infants with cystic fibrosis (PRESIS)
Mirjam Stahl (Heidelberg, Germany)

15:45 - 16:00
Use of computational fluid dynamics to model aerosol deposition in the lungs of patients with cystic fibrosis
Dearbhla Hull (Chichester, United Kingdom)

16:00 - 16:15
A randomised, controlled pilot study of sildenafil to treat exercise intolerance in moderate to severe cystic fibrosis
Jennifer Taylor-Cousar (Denver, United States)
Scientific Programme

Demographics of patients in a phase 2 trial of acebilustat in patients with cystic fibrosis (EMPIRE CF)
Stuart Elborn (London, United Kingdom)

16:15 - 16:30

Workshop
15:00 - 16:30

WS15, The right pill for everyone - advancing towards personalised therapy

Basic Science
Marcus Mall (Berlin, Germany)
Harry Heijerman (Utrecht, Netherlands)

From the bench to bedside: expanding CFTR modulators to rare mutations by response assessment in patients-derived materials and cellular models
Iris Silva (Lisboa, Portugal)
15:00 - 15:15

R334W CFTR, a severely compromised chloride conductance mutant, retains its bicarbonate conductance and responds to the corrector combination, C4+C18
Liudmila Cebotaru (Baltimore, United States)
15:15 - 15:30

Patterns of response to lumacaftor and ivacaftor in rectal organoids
Anabela S. Ramalho (Leuven, Belgium)
15:30 - 15:45

First functional characterisation of the R751L CFTR mutation using an ex vivo primary airway epithelial cell culture model
Iram Haq (Newcastle upon Tyne, United Kingdom)
15:45 - 16:00

Biomarkers to predict Orkambi efficacy: results of a prospective paediatric study
Alexandra Masson (Paris, France)
16:00 - 16:15

A phase 3, open-label study of tezacaftor/ivacaftor (TEZ/IVA) therapy: interim analysis of pooled safety, and efficacy in patients heterozygous for F508del-CFTR and a residual function mutation
Patrick Flume (Charleston, United States)
16:15 - 16:30

ECFS Tomorrow Session
15:15 - 16:15

MyCyFAPP: holistic system for children’s nutrition and PERT self-management
Carmen Ribes (Valencia, Spain)
Joaquim Calvo-Lerma (Valencia, Spain)
### WS16, Towards new markers of disease severity

**Immunology / Pulmonology / Inflammation**

Barry Plant (Cork, Ireland)
Charles Haworth (Cambridge, United Kingdom)

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
</tr>
</thead>
</table>
| 17:00 - 17:15 | **Early multi-dimensional assessment of Parameters to assess Response to Intra-Venous Antibiotic Treatment for pulmonary Exacerbations: The PRIVATE Study**  
Charlotte Addy (Belfast, United Kingdom) |
| 17:15 - 17:30 | **Serum levels of PAD4 auto-antibodies correlate with airway obstruction in cystic fibrosis patients: novel systemic marker of lung disease?**  
Ruchi Yadav (Athens, United States) |
| 17:30 - 17:45 | **Investigation of inflammatory and fibrotic biomarkers in the sputum of cystic fibrosis patients**  
Jan Christoph Thomassen (Cologne, Germany) |
| 17:45 - 18:00 | **Plasma YKL-40 levels and chitotriosidase activity in cystic fibrosis patients**  
Mina Hizal (Ankara, Turkey) |
| 18:00 - 18:15 | **Examining serum TH2 inflammatory exacerbation biomarkers in cystic fibrosis**  
Anna Siedlecki (VANCOUVER, Canada) |
| 18:15 - 18:30 | **Human epididymis protein 4 (HE4) plasma levels inversely correlate with improved FEV1 in cystic fibrosis patients under ivacaftor therapy as a new sensitive treatment efficacy biomarker**  
Béla Nagy Jr. (Debrecen, Hungary) |

### WS17, Unraveling the complexities of CFTR mutations and their impacts

**Genetics / Screening / Diagnosis**

Margarida Amaral (Lisbon, Portugal)
Eike De Wachter (Brussels, Belgium)

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
</tr>
</thead>
</table>
| 17:00 - 17:15 | **The multi-faceted nature of CFTR exonic mutations: impact on their functional classification**  
Caroline Raynal (Montpellier, France) |
| 17:15 - 17:30 | **Using a highly parallel sequencing assay for CFTR genotyping in ethnically diverse European patients with cystic fibrosis**  
Senne Cuyx (Leuven, Belgium) |
| 17:30 - 17:45 | **Functional characterisation and CFTR2 disease liability assignment of 48 missense variants**  
Karen Raraigh (Baltimore, United States) |
41st European Cystic Fibrosis Conference, 07 - 10 June 2017, Belgrade, Serbia

Scientific Programme

17:45 - 18:00
How organoid assay results concur with the clinical phenotype in an unusual patient with S1251N/G542X
Senne Cuyx (Leuven, Belgium)

18:00 - 18:15
Nasal potential difference measurement increases the diagnostic yield in patients with equivocal first-line cystic fibrosis investigations: the experience of a large national CFTR diagnostic service
Nicholas Simmonds (London, United Kingdom)

18:15 - 18:30
Characteristics of the reproductive system in adult male patients with cystic fibrosis
Svetlana Repina (Moscow, Russian Federation)

Workshop
17:00 - 18:30
Amphitheatre

WS18, Transplantation: waiting, caring, hoping

Open
Rachel Dunk (London, United Kingdom)
Lieven Dupont (Leuven, Belgium)

17:00 - 17:15
Waiting for lung transplant, manifold experiences: a literature review
Ulrika Skogeland (Stockholm, Sweden)

17:15 - 17:30
The transition towards lung transplant for cystic fibrosis in the United Kingdom
Natalie West (Baltimore, United States)

17:30 - 17:45
The introduction of a cystic fibrosis-specific advance care planning document
Fiona Cathcart (London, United Kingdom)

17:45 - 18:00
Evaluation of transplantation information delivered to patients and their relatives by the professionals from cystic fibrosis centres and transplant centres and from transplanted peer-patients in France
Valerie David (Nantes, France)

18:00 - 18:15
Does lung transplantation have an effect on the improvement of psychological status in adult patients with cystic fibrosis?
Ivana Lalic (Zagreb, Croatia)

18:15 - 18:30
Pregnancy after lung transplantation in cystic fibrosis patients
Frederikke Rönsholt (Copenhagen, Denmark)
WS19, Cystic Fibrosis-Related Diabetes: from diagnosis to pancreatic islet transplantation

Gastroenterology/Nutrition/Liver/Metabolic Complications

Isabelle Durieu (Lyon, France)
Helen White (Leeds, United Kingdom)

**Glucose abnormalities in Canadian and French cystic fibrosis patients: a Glyc-one database analysis**
Quitterie Reynaud (Lyon, France)
17:00 - 17:15

**The use of a questionnaire and continuous glucose monitoring to screen for hypoglycaemia in a cystic fibrosis clinic**
Natasha Armaghanian (Sydney, Australia)
17:15 - 17:30

**Early screening and treatment of paediatric Cystic Fibrosis-Related Diabetes (CFRD) slows respiratory decline**
Charlotte Walker (London, United Kingdom)
17:30 - 17:45

**Glucose and insulin area under the curve (AUC) can differentiate between cystic fibrosis patients that may benefit from early insulin treatment**
Liora Soesman (Jerusalem, Israel)
17:45 - 18:00

**The main mechanism associated with glucose intolerance in older patients with cystic fibrosis is insulin resistance and not progressive insulin secretion deficit**
Valerie Boudreau (Montreal, Canada)
18:00 - 18:15

**Feasibility and efficacy of combined lung and pancreatic islet transplantation in Cystic Fibrosis-Related Diabetes: a pilot study**
Laurence Kessler (Strasbourg, France)
18:15 - 18:30

WS20, Late Breaking science

Latebreaking Science

Harm Tiddens (Rotterdam, Netherlands)
Damian Downey (Belfast, United Kingdom)

**The importance of early diagnosis of Mycobacterium abscessus complex**
Tavs Qvist (Copenhagen, Denmark)
17:00 - 17:23

**Evaluation of ELX-02 in Cystic Fibrosis Organoids with Non-Sense Mutations**
Pedro Huertas (Waltham, United States)
17:23 - 17:38

**A double-blind, placebo-controlled, randomised study of SPX-101, a novel modulator of ENaC cell surface density in adults with cystic fibrosis.**
Isabelle Fajac (Paris, France)
17:38 - 17:53
Scientific Programme

17:53 - 18:03
**BEAT-CF: An adaptive trial design for the treatment of respiratory tract exacerbations**
André Schultz (Perth, Australia)

18:03 - 18:13
**Panel discussion: Opportunities to run more effective clinical trials**
Silke van Koningsbruggen-Rietschel (Cologne, Germany)
Stuart Elborn (London, United Kingdom)
André Schultz (Perth, Australia)

Interactive Poster Discussions
17:00 - 18:30

**IPD2, What do we learn from CFTR modulator use in real life**
Open
Jane Davies (London, United Kingdom)
Kris De Boeck (Leuven, Belgium)

- Disease progression in patients with cystic fibrosis treated with ivacaftor: analyses of real-world data from the US and UK cystic fibrosis Registries
  Nataliya Volkova (Boston, United States)

- Real-world outcomes in patients with cystic fibrosis treated with ivacaftor: 2016 US and UK cystic fibrosis Registry analyses
  Nataliya Volkova (Boston, United States)

- The effects of 3-year ivacaftor use on lung function and intravenous days seen in UK cystic fibrosis Registry data
  Simon J Newsome (London, United Kingdom)

- Long-term effects of ivacaftor in patients with G551D mutation and mild lung disease
  Helmut Ellemunter (Innsbruck, Austria)

- Effects of ivacaftor in patients with cystic fibrosis and severe lung disease carrying CFTR mutations with residual function
  Donatello Salvatore (Potenza, Italy)

- Ivacaftor therapy in patients with severe baseline lung disease carrying a residual function mutation
  Ruth Mitchell (Manchester, United Kingdom)

- Retrospective observational study in cystic fibrosis patients homozygous for F508del treated with lumacaftor/ivacaftor in a compassionate use programme
  Paola Iacotucci (Naples, Italy)

- Dose modification of lumacaftor/ivacaftor and the immediate effects on lung function in cystic fibrosis patients with advanced lung disease and 12-month outcomes in this cohort
  Natalia Popowicz (Perth, Australia)

- Lumacaftor/ivacaftor improves 6 minute walk test distance, FEV1 and reduces exacerbations in severe cystic fibrosis lung disease
  Theeba Thiruchelvam (New Lambton NSW, Australia)
Scientific Programme

Observational study of glucose tolerance abnormalities in patients with cystic fibrosis homozygous for Phe508del CFTR treated by lumacaftor-ivacaftor
Bastien Misgault (Strasbourg, France)

The potentially beneficial CNS-activity profile of ivacaftor and its metabolites
Elena Schneider (Melbourne, Australia)

Treatment with Orkambi™ in Phe508del homozygous cystic fibrosis patients is associated with improvement in cognition
John Wilson (Melbourne, Australia)

Treatment with ivacaftor in cystic fibrosis patients with the G551D mutation is associated with improvement in cognition
John Wilson (Melbourne, Australia)
Saturday, 09 June 2018

**Symposium**
09:00 - 10:30

**Blue Hall**

**S25, Where are we with novel, single-drug approaches to CF therapy?**

Upon completion of this session participants should be able to:
- Describe the molecular mechanisms that can be exploited to repair CFTR at the DNA and RNA levels
- Explain the concept of CFTR amplifier as an adjuvant therapeutic approach in cystic fibrosis
- Evaluate the importance of modifier genes on CFTR function

Dorota Sands (Warsaw, Poland)
George Z. Retsch-Bogart (Chapel Hill, United States)

- **Gene editing**
  Patrick T. Harrison (Cork, Ireland)
  09:00 - 09:22

- **mRNA repair**
  Batsheva Kerem (Jerusalem, Israel)
  09:22 - 09:44

- **Alternative chloride channels**
  Karl Kunzelmann (Regensburg, Germany)
  09:44 - 10:06

- **Targeting ENaC**
  Rob Tarran (Chapel Hill, United States)
  10:06 - 10:30

**Hall 1**

**S26, The new age of inflammation in cystic fibrosis**

Upon completion of this session participants should be able to:
- Get an overview of inflammation in CF
- Understand the pros and cons of treating inflammation in CF
- Learn about possible future anti-inflammatory therapies in CF

Stuart Elborn (London, United Kingdom)
Scott Bell (Brisbane, Australia)

- **Systemic airway and gut inflammation in cystic fibrosis**
  Burkhard Tümmler (Hanover, Germany)
  09:00 - 09:22

- **Inflammation should be treated in cystic fibrosis - PRO**
  Jerry A. Nick (Denver, United States)
  09:22 - 09:40

- **Inflammation should be treated in cystic fibrosis - CON**
  Scott Bell (Brisbane, Australia)
  09:40 - 09:58

- **Discussion**
  09:58 - 10:08

- **Pipeline of anti-inflammatory treatments**
  Stuart Elborn (London, United Kingdom)
  10:08 - 10:30
Scientific Programme

Symposium
09:00 - 10:30

S27, Co-evolution of CF pathogens - do they drive disease?

The purpose of this session is to:
- Give clinicians and basic scientists an understanding how multispecies interactions within airway microbial communities are critical for airway colonisation, responses to perturbations, and transitions between health and disease
- Discuss the role of multispecies social interactions in shaping *P. aeruginosa* pathogenicity in the CF lung with a particular focus on *Staphylococcus, Streptococcus* and *Aspergillus* species

Niels Høiby (Copenhagen, Denmark)
Cristina Cigana (Milan, Italy)

- **Microbiota interactions - understanding the networks**
  09:00 - 09:22
  Michael Tunney (Belfast, United Kingdom)

- **Pseudomonas-Staphylococcus interactions**
  09:22 - 09:44
  Christian van Delden (Geneva, Switzerland)

- **Aspergillus and Pseudomonas**
  09:44 - 10:06
  Craig Williams (Glasgow, United Kingdom)

- **Pseudomonas and Streptococcus**
  10:06 - 10:30
  Michael G. Surette (Calgary, Canada)

Symposium
09:00 - 10:30

S28, Important aspects of GI and nutrition in adult CF care

Upon completion of this session participants should be able to:
- Learn the gastro-intestinal aspects of an adult CF patient
- Learn the impact of nutrition in post transplantation care

Helen White (Leeds, United Kingdom)
Michael Wilshanski (Jerusalem, Israel)

- **Adult-specific gastro-intestinal complications - overview**
  09:00 - 09:44
  Chee Y. Ooi (Randwick, Australia)

- **Cystic fibrosis and disorders of the large intestine: colorectal cancer**
  09:44 - 10:06
  James Abraham (Minneapolis, United States)

- **Nutritional care after transplantation**
  10:06 - 10:30
  Francis Hollander (Utrecht, Netherlands)
S29, A new era: Registry-based clinical studies

Upon completion of this session participants should be able to:
- Appraise the value of Registries to design time and cost-effective clinical studies
- Identify how CF Registries can answer fundamental questions that might otherwise go unanswered
- Discover how contemporary comparison groups can be construed in CF Registries

Lutz Nährlich (Giessen, Germany)
Ed McKone (Dublin, Ireland)

Establishing European guidelines for Registry-based studies 09:00 - 09:22
Lutz Nährlich (Giessen, Germany)

Real-world outcomes of Registry-based Pharmacovigilance 09:22 - 09:44
Siobhan Carr (London, United Kingdom)

Registry studies: the feasibility of encounter based vs. yearly data collection 09:44 - 10:06
Kevin Southern (Liverpool, United Kingdom)

Clinical trials and the Cystic Fibrosis Foundation Patient Registry 10:06 - 10:30
Margaret Rosenfeld (Seattle, United States)

S30, Mucus: barrier properties and interactions

Upon completion of this session participants should be able to:
- Identify how mucins contribute to the barrier function of mucus and the challenges this poses for drug delivery
- Remember how CF pathogens can disrupt mucus properties
- Discuss how mucins inhibit biofilm formation and how mucus affects immune defence of the lung

Nicoletta Pedemonte (Genoa, Italy)
Dave Thornton (Manchester, United Kingdom)

The role of mucins and mucus clearance in the barrier properties of the airways 09:00 - 09:22
Brian Button (Chapel Hill, United States)

The effect of mucin on Pseudomonas biofilm formation 09:22 - 09:44
Katharina Ribbeck (Cambridge, United States)

The interplay between the mucus barrier and Aspergillus fumigatus 09:44 - 10:06
Dave Thornton (Manchester, United Kingdom)

Study of mucus barrier formation using Muc5b-GFP mouse 10:06 - 10:30
Jean-Luc Desseyn (Lille, France)
Scientific Programme

Closing Plenary
11:00 - 12:30
Blue Hall

Closing Plenary - Bringing new therapies to all patients
Isabelle Fajac (Paris, France)
Kris De Boeck (Leuven, Belgium)
Predrag Minic (Belgrade, Serbia)

The future of precision medicine 11:00 - 11:30
Kors Van der Ent (Utrecht, Netherlands)

Overcoming the obstacles posed by reimbursement issues to bring drugs 11:30 - 12:00
to treat rare diseases to patients: challenges for cystic fibrosis
Ron Akehurst (Sheffield, United Kingdom)

ECFS President Address 12:00 - 12:30
Isabelle Fajac (Paris, France)

Closing Ceremony
12:30 - 13:00
Blue Hall

Closing Ceremony