

Letter from the President

Dear Friends,

In these unprecedented times with the COVID-19 pandemic, we hope that you and your dear ones are all safe and well.

As you already know the Basic Science Conference has been cancelled and rescheduled for 17-21 March 2021 in Portugal. Carlos Farinha, Nicoletta Pedemonte and Jeff Brodsky kindly accepted to chair next year's Conference. I take this opportunity to heartily thank them for all their efforts and commitment over the last months: we had difficult decisions to take and the best for the conference and participants were always at the forefront of their thoughts and guiding their advice. In the coming months, the programme will be carefully reviewed and updated if and when needed.

Understandably, the Annual Conference in Lyon which was planned in early June will not take place. We have to cancel our annual face-to-face meeting and postpone the burst of lively and fruitful discussions and educational activities this conference usually allows, but we are happy to announce the organisation of a digital mini-43rd European CF conference in the fourth week of September. It will include excerpts of the Scientific Programme which had been prepared for Lyon. We have already 8 symposia confirmed for 24 and 25 September and we will keep you informed of the programme as it develops, but already mark your calendar not to miss the event! We look forward to seeing you online in large numbers.

Quite a few ECFS Working Groups and projects have also decided to keep their planned meetings and will hold online sessions and meetings over the dates of the conference next week or in September. Please see further on in this newsletter the list of coming meetings. In the meantime, we are happy to already confirm that the next Annual Conference will be organised in Milan, Italy, on 9-12 June 2021.

As part of our commitment to support young researchers, we granted the Gerd Döring award to Dr. Paolo Scudieri to honour his early career as a scientist and three Young Investigators have been awarded based on the scientific merit of their work submitted to the conference. You will find a short presentation of this year's awardees and their research later on in this newsletter. We will unfortunately not be able to meet them in Lyon but will honour them in person at the conference in Milan next year.

We are all disappointed by the cancellation and postponement of our conferences but I am happy to see the energy and enthusiasm of all of you to keep our community as lively as possible.

Your feedback is also crucial to us and I invite all ECFS electorate to take part in the elections for Board members and ECFS Presidency that opened on May 20th. You will learn more about these elections in the newsletter.

Keep well,

Isabelle Fajac



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ECFS Board Elections

The ECFS is holding elections for 3 Board positions: ECFS President Elect and 2 Ordinary Board Members.

Marcus Mall and Kevin Southern will end their terms in June 2020 and Isabelle Fajac will finish her term as President in 2021.

We have been lucky to receive applications for ECFS Board member from five highly valued senior figures of the CF community with international reputation.

Candidates' information and links to supporting documents such as the candidates' nominations, application letters and CVs are available [here](#). Invitations to take part in the elections have been sent to the ECFS electorate by email and access to the online secured voting poll can be found [here](#)

Job Descriptions are available for both the President and Board Members [President's Job Description](#) | [Board Member's Job Description](#).



Charles Haworth (UK)



Dorota Sands (PL)



Carsten Schwarz (DE)



David Sheppard (UK)



Michal Shteinberg (IL)

Upcoming Events

- **ECFS Board Summer Meeting**
2 June 2020
- **Annual General Meeting ECFS**
5 June 2020
- **ERS Virtual Congress**
7-8 September 2020
- **ECFS Digital Conference**
23-25 September 2020
- **North American Cystic Fibrosis Conference**
22-24 October 2020
Phoenix, AZ (US)
- **ECFS Winter Meetings**
20 –22 January 2021
Brussels
- **ECFS Diagnostic Network WG Meeting,**
February 2021
- **Basic Science Conference,**
17-21 March 2021
Albufeira, Portugal
- **43rd European CF Conference,**
9-12 June 2021
Milan, Italy

ECFS Annual General Meeting

Save the date!

The Annual General Meeting will be held on Friday 5 June 2020 from 17:00 to 18:00 CET.

As the conference in Lyon has been cancelled, the Annual General Meeting will be organised online. We need your active participation and look forward to a great discussion. The link to the meeting will be sent shortly by email.

Upcoming Meetings

- **ECFS Exercise Working Group Meeting**
3 June 12:00-15:00 CET
- **ECFS CTN Steering Committee Meeting**
3 June 13:00-14:45 CET
- **ECFS CTN Training Meeting**
3 June 15:00-16:30 CET
- **Webinar: We are screening... now what? Different treatment modalities in CF mental health**
4 June 12:45-13:45 CET
- **ECFSPR Scientific Committee Meeting**
4 June 13:00-14:30 CET
- **ECFS Standards of Care Group Meeting**
4 June 13:30-14:30 CET
- **International Physiotherapy Group for Cystic Fibrosis Annual General Meeting**
4 June 13:45-14:45 CET
- **ECFS Mental Health Working Group Meeting**
4 June 15:30-17:00 CET
- **ECFS Patient Registry Steering Group Meeting**
5 June 13:30-15:00 CET

AGENDA

1. President's Report – Isabelle Fajac
2. Secretary's Report – Isabelle Sermet
3. Treasurer's Report – Kevin Southern
4. ECFS Board Elections – Isabelle Fajac
5. Update on Journal of Cystic Fibrosis – Patrick Flume
6. CTN Report – Silke van Koningsbruggen-Rietschel
7. ECFS Standards of Care Report – Carlo Castellani
8. ECFS Patient Registry Report – Lutz Naehrlich / Andreas Jung
9. ECFS Education Report – Daniel Peckham
10. ECFS Working Group Reports
 - Antimicrobial Resistance Working Group
 - Basic Science Working Group
 - Diagnostic Network Working Group
 - Exercise Working Group
 - Fungal Pathogens Working Group
 - Mental Health Working Group
 - Neonatal Screening Working Group
 - Pulmonary Exacerbations Working Group
11. ECFS Special Interest Groups
 - Nursing Special Interest Group
 - European CF Pharmacy Group
 - European CF Nutrition Group
 - European Psychosocial Special Interest Group
12. Presentation of the 2021 European CF Conference

Digital mini-43rd European CF Conference

Mark your calendar!

ECFS e-Conference launching on the 4th week of September!
During week 39 (between 21st and 25th September), we will organise an online, mini 43rd European CF conference.
We currently have 8 sessions confirmed on 24 and 25 September.
We will keep you informed as the programme develops.



List of symposia planned for 24-25 September 2020

- *Following treatment outcomes in patients on CFTR modulators*
- *The future of clinical research*
- *Non-Pseudomonas aeruginosa pathogens in cystic fibrosis*
- *New challenges for diagnosis in cystic fibrosis, from pregnancy to adulthood*
- *Body and Soul Psychological influences on physical outcomes*
- *From disease mechanisms to novel therapies*
- *Updates for the gut and the pancreas.*

ECFS2020 - Abstracts

The ECFS will publish the abstracts submitted to the 43rd European Cystic Fibrosis conference in June as a supplement to the Journal of Cystic Fibrosis. Abstracts will also be available on a platform accessible to ECFS members and all registered participants to the Lyon conference and the September mini-conference.

Gerd Döring Award 2020

The Gerd Döring Award is an initiative of the European Cystic Fibrosis Society and is given annually to honour an exceptional young European scientist in their early career. The Award includes a monetary donation of 5,000 euro to support research. This year, the Award is given to Paolo Scudieri in acknowledgement of his work.

Paolo Scudieri



Paolo Scudieri has a Master degree in Biological Sciences and in 2018 he obtained a PhD in Paediatric Sciences - Genetics from the University of Genoa. He has worked in the laboratory of Prof. Luis Galiotta, previously at the Istituto Giannina Gaslini (Genoa) and since 2016 at the Telethon Institute of Genetics and Medicine (TIGEM) (Pozzuoli). His research has been mainly dedicated to understand the role and the structure-activity relationship of the TMEM16A channel, to study the molecular and functional modifications of ion transport associated with mucous cell hyperplasia in the bronchial epithelium, and to investigate the role of bicarbonate and pH in cystic fibrosis basic defect.

His paper, *Increased expression of ATP12A proton pump in cystic fibrosis airways* (Scudieri et al., JCI Insight 3(20):e123616, 2018), describes interesting findings regarding the expression and function of ATP12A, which may have important implications in the understanding of the role of this protein as a therapeutic target in cystic fibrosis.

The study reports for the first time that the airways of CF patients have enhanced expression of ATP12A proton pump. This phenomenon, which may arise from infection and inflammation, probably contributes to lung disease by acidification of the airway surface. This study also solves an important controversial issue regarding the trafficking of ATP12A. Paolo found that ATP1B1 is the subunit required by ATP12A to traffic

ECFS 2020 – Young Investigators Award

Following the recommendations of the Steering Committee during the abstract review process of the Annual conference, the ECFS wishes to commend the quality of the work presented in the abstracts of some young investigators under the age of 35 having applied for the award. The Young Investigator Award includes a monetary grant of 750 €, a free registration to next year's conference and a 2020 ECFS membership subscription.

Frederik Holz



Frederik Holz received his medical training in Munster and Berlin. He holds a doctoral scholarship from the Hans-Böckler-Foundation. In 2017, Frederik has started a pediatric residency at Charité University hospital in Berlin.

His research focusses on the phenomenon of arthropathy in patients with CF (CFA). It is a common co-morbidity that can significantly impact the quality of life. Even though CFA is gaining more importance due to the rising life expectancy of CF patients, it remains difficult to state a clear definition since the causes seem to be multifactorial and the pathogenesis is not fully understood yet.

The aim was to assess the clinical manifestation of CFA based on radiological findings, clinical symptoms and scores gathered in a prospective study.

With this research different stages for the clinical manifestation of CFA can now be described which may lead to a better understanding of the clinical phenotype of the disease and its different forms and intensities. Subsequently these findings may contribute to further improvement in the process of diagnosis and assessment as well as CFA therapy.

Eline Lauwers



After her master in Physiotherapy and Rehabilitation Sciences, Eline Lauwers started her PhD at the Laboratory for Experimental Medicine and Paediatrics at the University of Antwerp in Belgium. Her research explores novel outcome measures for the evaluation of respiratory therapy in children with obstructive lung diseases. More specifically, she focusses on digital lung auscultation and functional respiratory imaging. She believes that these innovative techniques will contribute to the development of improved targeted therapies for patients with cystic fibrosis.

Her recent research involves the use of Functional Respiratory Imaging (FRI) to evaluate the short-term effects of Orkambi (lumacaftor/ivacaftor). Orkambi has shown modest benefits in previous studies, but the exact effects remain unclear. In order to get a more in-depth understanding of the treatment effects, FRI was performed. This innovative approach combines CT imaging with computational fluid dynamics, providing detailed information about the lung structure and function. The results showed that, after three months of treatment with Orkambi, lung hyperinflation and airway geometry significantly decreased in patients homozygous F508del aged ≥ 12 years. Both air-trapping and the decrease in airway geometry were correlated with an improvement in quality of life. Consistent with previous research, lung function parameters measured by spirometry showed limited changes. These results indicate that the FRI parameters, reflecting regional and distal lung structures, are more sensitive to changes caused by Orkambi than conventional spirometry.

Alessandra Murabito



Alessandra Murabito received her Master Degree in Molecular Biotechnology in 2017 at the University of Torino, where she characterized a PI3K γ mimetic peptide as a bronchodilator and anti-inflammatory drug in respiratory diseases in the laboratory of Professor Hirsch, a leading expert in PI3K signaling. Under the guidance of Prof. Alessandra Ghigo, Alessandra Murabito began her Ph.D. which is focused on the role of PI3K γ in epithelial cells and the use of the PI3K γ mimetic peptide for the treatment of cystic fibrosis.

In the last years, her research has focused on the development of a peptide inhibiting the scaffold function of PI3K γ (Patent WO/2016/103176), which negatively regulates cAMP. Her work demonstrated that the compound targets several pathological aspects in cystic fibrosis, promoting smooth muscle relaxation and preventing lung neutrophilia. Now the focus is set on the research on epithelial cells, given the importance of cAMP on CFTR regulation. It already showed that the peptide stimulates CFTR phosphorylation and gating, and synergizes with Orkambi $\text{\textcircled{C}}$. Moreover, the compound increases the amount of CFTR at the plasma membrane. The current aim is to try to unravel how PI3K γ controls CFTR stability, since preliminary data indicate that it elevates the core-glycosylated F508del-CFTR and so the possible involvement of the unconventional secretory pathway. Overall, these results demonstrate that the PI3K γ -peptide may be exploited therapeutically to provide bronchodilation, anti-inflammatory effects and CFTR modulation.

ECFS / CF Europe Post-Doctoral Research Fellowship



With the aim to foster basic and translational research on CF, the ECFS and CF Europe organised a call for applications for their Post-Doctoral Research Fellowship programme. We received eleven applications. Successful applications after a first check for compliance were sent for external review and the ECFS Selection Committee gave recommendations to the Boards of the ECFS and CFE for the successful candidates. We thank the Selection Committee composed of Isabelle Fajac (FR), Carlos Farinha (PT), Mike Gray (UK), Sylvia Hafkemeyer (DE), Claus Moser (DK) and Nicoletta Pedemonte (IT) for their enthusiasm and commitment.

Three candidates have been selected for a two-year post-doctoral research fellowship. The names of the ECFS / CFE Post-doctoral research fellows will be announced at the ECFS AGM on 5 June. Please join us at the AGM to congratulate them!

COVID-CF project in Europe

COVID-19 IN PEOPLE WITH CF IN EUROPE

Data up to 20 May 2020



Background

CF patient registries throughout Europe are collecting data about people with CF who become infected with SARS-Cov-2, causing the illness COVID-19.

Countries that contribute annual data to the ECFS Patient Registry (ECFSPR) were invited to report COVID-19 case data of people with a confirmed diagnosis of CF. Here we present a weekly update of centralised, anonymised data.

The data is preliminary, incomplete, might change over time, and the number of cases is low. Therefore this information should not be used to direct clinical decisions.

Covid-19/CF cases



12 countries with cases
65 cases reported
56 cases documented

Status of COVID-19 illness



3 died
13 ongoing
32 recovered
8 unknown

Gender



27 male
29 female

Age



14 <18 years
35 18-49 years
3 >50 years

Severity



27 mild cases
7 severe cases
3 critical cases

Further information

- 41 patients were hospitalised (6 in the ICU) and 2 patients needed ventilation
- 11 patients were asymptomatic
- The top 5 symptoms were fever, increased cough, fatigue, increased sputum production and increased breathlessness.

CF patient registries throughout Europe have collected data about people with CF who become infected with SARS-Cov-2.

An update is posted every week on the ECFS website. The data is preliminary and incomplete. An in-depth statistical analysis will be performed and published as soon as possible.

Last update 20 May 2020

[Read more](#)

A day in the life of...

At ECFS we take pride in our diverse membership which is so important in the advancement of CF and in supporting rich discussions at our various meetings and Conferences. Although the Society is based in Europe, we warmly welcome members from many other parts of the world. For example, this year almost 10% of our members come from North America, over 3% from Australasia/Oceania and 2% from Asia. Our members also have many different job titles including working as doctors, allied health professionals, lecturers and lab technicians to name a few. This is why we would like to hear from you and find out what a typical day in the life of our members is like! What does your typical day look like? Do you work in a clinical or laboratory setting? Are you involved in any special projects or research? What made you want to work in the field of CF? What are the particularities of working in your country? We will then feature this snap-shot in subsequent Newsletters to highlight our diverse Society. Please send all pieces alongside a headshot to: membership@ecfs.eu

We look forward to hearing from you soon!

Current References in Cystic Fibrosis

Many thanks to Dr. Henry Ryley for compiling the current references in Cystic Fibrosis that you can download through this [link](#).

Please contact us if you have news items you would like to have included in future Newsletters or published on our website!

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A busy week ahead....

Date	Online Meeting / Webinar	Start	End
02/06/2020	ECFS Board Meeting	11:00	13:00
02/06/2020	ECFS Patient Registry Executive Committee Meeting	13:30	14:30
02/06/2020	ECFS Board Meeting	15:00	17:00
03/06/2020	ECFS Exercise Working Group Meeting	12:00	15:00
03/06/2020	ECFS CTN Steering Committee Meeting	13:00	14:45
03/06/2020	ECFS CTN Training Meeting	15:00	16:30
04/06/2020	Webinar: We are screening...now what? Different treatment modalities in CF mental health	12:45	13:45
04/06/2020	ECFSPR Scientific Committee Meeting	13:00	14:30
04/06/2020	ECFS Standards of Care Group Meeting	13:30	14:30
04/06/2020	International Physiotherapy Group for Cystic Fibrosis (IPG/CF) Annual General Meeting	13:45	14:45
04/06/2020	ECFSPR Pharmacovigilance Working Group Meeting	15:00	16:30
04/06/2020	ECFS Mental Health Working Group Meeting	15:30	17:00
04/06/2020	ECFSPR Annual Report Working Group	17:00	18:00
05/06/2020	ECFSPR Patient Projects & Social Media Meeting	09:00	10:00
05/06/2020	European Registry Steering Group Meeting	13:30	15:00
05/06/2020	ECFS Annual General Meeting	17:00	18:00