



ECFS NEWSLETTER - Issue 48 -May 2015

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Letter from the President and President-Elect

Dear Friends,

There has been a lot of activity at the ECFS since the last edition of our newsletter and here is a brief review of the special projects, what's new with our society and a few highlights of the upcoming 38th European Cystic Fibrosis Conference taking place in just a few weeks, this year in Brussels, Belgium.

Changes in the ECFS Board are already happening. Kris De Boeck as President Elect, has taken on the role of President and will officially start her 3 year mandate as ECFS President this June. Also, Stuart Elborn, Dominique Hubert and Milan Macek will be leaving the ECFS Board. On behalf of the ECFS, we extend our thanks for their excellent contributions; it has been a pleasure working with them. The results of the elections held this month will be presented during the Annual General Assembly (AGM) in Brussels. We would like to thank all who have submitted their vote and we very much hope that as many of you as possible will attend the AGM. We value your involvement and it is really important to have members contribute to elections and input new ideas. More information on the AGM will appear further down in the newsletter.

In January, the **ECFS Clinical Trials Network** and the **ECFS Patient Registry** held their Steering Group meetings in Barcelona, Spain. With the two groups meeting in parallel, there was an excellent opportunity for all to meet, discuss common subjects and review ways of cooperation. Both the CTN and ECFSPR remain key projects of the Society. The support of several national organisations continues to be greatly appreciated and we look forward to working closely with them and increasing our partnerships in the future.

The **12th ECFS Diagnostic Network Working Group Meeting** took place in Warsaw, Poland, 12-14 February 2015, hosted by Dorota Sands. An important part of the DNWG meetings is the contribution of Young Investigators. We were able to support Young Investigators (under 35 years of age) selected for their scientific work through Travel Grants (max 250€). Young Investigators from Italy, Poland and Germany presented their work during the meeting. The full meeting report is available on the ECFS website [here](#).

The **ECFS Basic Science Conference** was held in Albufeira, Portugal, 25-28 March. With a record number of abstracts and of attendees, the conference was a resounding success. We would like to thank the organizers, Marc Chanson (CH), Fred Becq (FR) and Martina Gentzsch (US) for putting together a high quality programme. The [abstract book](#) is available on the ECFS website.

Finally, we are happy to announce that the second Edition of the ECFS Book, **Living Longer with Cystic Fibrosis**, will be available at the Brussels conference. Many thanks go to Dominique Hubert and Nicholas Simmonds, Editors of the book for their hard work. We hope that this book will help to attract more adult care physicians to the field of CF. The book will be offered to all ECFS members.

We are rapidly approaching our summer conference in Brussels and the final preparations are well under way. The Scientific Committee has put together an outstanding programme and we have some exciting new basic research and results from clinical trials to discuss. You can find the [final programme](#) on the ECFS website. We would like to thank the Scientific Committee and the Steering Committee for all their hard work over the last year in preparing the programme, selecting the abstracts and sorting the various sessions. Besides the conference Smartphone Application featuring the programme and other useful information, other recent initiatives include the Interactive Case studies session, some morning 'meet the experts' sessions and the ECFS Tomorrow lounge aimed at young investigators interested in building their career in Cystic Fibrosis. You can find more about the Interactive sessions and the ECFS Tomorrow further in this newsletter.

This year, we are also running a one day Quality management/Quality Improvement course on 09 June and a one day CF Course on 10 June. Both courses have been awarded 6 credit points for continuing medical education.

As part of our commitment to support young researchers, three Young Investigators Awards and 21 travel grants have been awarded to young investigators based on the merit of their abstract submissions.

Please don't miss a visit to the "CF Community" booth area in the Exhibition Hall for information on the various initiatives and programmes. There will be representatives from the ECFS, the ECFS-Clinical Trials Network, the ECFS Patient Registry, Elsevier (JCF Publisher), CF Europe, The Belgian CF association and CF Worldwide.

Many thanks, as always, to Dr. Henry Ryley for compiling the current references in Cystic Fibrosis contained at the end of this Newsletter.

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

Yours sincerely,

Stuart Elborn, ECFS President

Kris De Boeck, ECFS President Elect

12th ECFS Basic Science Conference - Albufeira, Portugal

The **ECFS Basic Science Conference** was held in Albufeira, Portugal between 25-28 March. With a record number of abstracts and of attendees (close to 200), the conference was a resounding success. Grateful thanks to the organisers, Marc Chanson (CH), Frédéric Becq (FR) and Martina Gentsch (US) for putting together a high quality programme. The [abstract book](#) is available on the ECFS website.



At the ECFS Conference in Brussels, Frédéric Becq will present the Highlights of the 12th ECFS Basic Science Conference in the Late Breaking Science session (WS22 – Friday 12 June - 17:00-18:30)

ECFS Tomorrow



Last year we launched the “ECFS Tomorrow” initiative in Gothenburg. We received a positive feedback and we are happy to prepare its second edition in Brussels.

We see the ECFS Tomorrow as a means to improve one of the fundamental objectives of the society – to support and engage young scientists working in the field of Cystic Fibrosis.

It is essential to provide professionals who are new to the community with opportunities to connect in a meaningful way with current leaders and experts. In turn, the new generation of specialists bring up-to-date approaches and practices to the wealth of experience that already exists within the society’s ranks. Over the four days of conference, people from our multidisciplinary community researching and treating Cystic Fibrosis will come together at the ECFS Tomorrow Lounge to exchange ideas, partake in the ECFS Lounge Programme or simply take a moment to pause and reflect on the themes of the day.

Two workshops will be given at the Lounge

On Thursday night, we will also organise a special ECFS tomorrow networking event outside the conference centre. In a traditional Belgian beer pub, teams will have the possibility to play a Quiz on CF and ... on Beer and Belgium! The prize for the winner will be a free registration to the ECFS Conference in Basel next year. At 10 pm there will be a performance of rock/blues band Sticky Fingers which was initially co-founded by Harm Tiddens, with special guest Dominik Hartl on guitar! Not to be missed!

Cystic Fibrosis Courses

- A one day CF Course is organised in Brussels on Wednesday 10 June, the day of the Opening of the 38th European CF Conference.

The course is intended for early career medical and paramedical health care workers involved in CF care and /or CF research. This course has been granted 6 CME credits by the European Accreditation Council for Continuing Medical Education (EACCME).

Click [here](#) for more details on the programme.

- The ERS and the ECFS will jointly organise a course on Cystic Fibrosis in Prague, Czech Republic, on 19-21 November 2015.

This course approaches the disease of CF from a multi-disciplinary and collaborative approach. The important topics of epidemiology, diagnosis, pathophysiology of CF, treatment, and psychological issues are covered, as well as the key factors involved in the organisation of CF care. Two specific tracks will be available for paediatric and adult care.

This course has been granted 17 CME credits by the European Accreditation Council for Continuing Medical Education (EACCME). The number of credits that you will receive corresponds to your attendance during the course.

Click [here](#) for more details on the programme.

Quality Management/ Quality Improvement Course

Education is one of the major objectives for the ECFS. This year a one day course on Quality Management/Quality Improvement is organised in Brussels on Tuesday 9 June.

The course is intended for CF centre directors and co-workers from all European countries with a special interest in quality management/quality improvement tools. This course has been granted 6 CME credits by the European Accreditation Council for Continuing Medical Education (EACCME).

ECFS Award 2015

The ECFS Award is given annually to honour a person who has made an outstanding contribution to our basic understanding of Cystic Fibrosis or to the treatment or care of patients with Cystic Fibrosis.

This year the award goes to Dr. Carlo Castellani. The ECFS wishes to recognise and honour the significant contribution of Dr. Castellani to neonatal screening and the genetic diagnosis of people with Cystic Fibrosis

and to the European Cystic Fibrosis Society. As an ECFS Board member, he led some key consensus conferences which have led to highly cited consensus statements on neonatal screening and genetic diagnosis. Most recently, he superbly led the Standards of Care Consensus which was an enormous piece of work likely to impact on CF care throughout Europe.



Gerd Döring Award

Professor Gerd Döring was an outstanding leader of the ECFS. During his mandates as ECFS President, he initiated a highly successful annual conference on basic science and consensus conferences on the treatment of lung infection, early intervention and prevention of lung disease, drug inhalation and related devices, nutrition, along with recommendations for several clinical trials in Cystic Fibrosis, which have had a significant impact on treatment.

The support of young investigators was a high priority for him and to honour his enormous contribution, the ECFS has initiated a Gerd Döring Award that will be given annually to honour an exceptional early career young European scientist.

The award is primarily judged on a paper published in the previous 3 calendar years, which has made a significant impact on the understanding or treatment of Cystic Fibrosis. This year, the Gerd Döring Award has been granted to Ms Florijn Dekkers, in acknowledgement of her recent remarkable contribution to Cystic Fibrosis. The Award, which includes a monetary grant of 5.000 Euro to support research, will be presented during the Opening Plenary of the Brussels Conference on 10 June.

Florijn Dekkers started her PhD project in 2010 in Jeffrey Beekman's lab in Utrecht, The Netherlands. She rapidly learned to culture intestinal organoids, a stem cell-based culture technology that allows rapid and limitless expansion of subject-specific tissue. Within 18 months of the observation, Nature Medicine accepted her first paper describing forskolin induced swelling of intestinal organoids as disease model for CF. This was the first disease model described in adult stem cultures with great impact on CF diagnosis, drug discovery and personalized medicine. This paper described a novel functional CFTR assay in organoids generated from rectal tissue biopsies that were leftovers from diagnostics. This simple and robust assay relies on CFTR-dependent phenotypic organoid swelling induced by forskolin, and can be used to quantitate the residual subject-specific CFTR function and response to CFTR-modulating drugs.

Importantly, organoids from individuals with either different or identical CFTR mutations responded differently to medication.

Florijn will present her research in the Late Breaking Science Session, Friday, June 12, 17:00 – 18:30 in Hall 100.



ECFS Young Investigators Awards

Following the recommendations of the Scientific Committee during the abstracts review process, 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 the Conference, and a 2015 ECFS membership subscription.

We wish to extend our congratulations to the following Young Investigators:



Susanne Dittrich

Susanne Dittrich is a clinician scientist at the Translational Lung Research Center Heidelberg (TLRC), member of the German Center for Lung Research (DZL) and the Molecular Medicine Partnership Unit (MMPU), University of Heidelberg. In 2013, she graduated from medical school at the Dresden University of Technology, where she conducted her experimental M.D. thesis on pulmonary inflammation in acute respiratory distress syndrome. This was associated with a research stay at the Massachusetts General Hospital, Boston (USA). In 2015, Susanne Dittrich was awarded with the HRCMM (Heidelberg Research Center for Molecular Medicine) Career Development Fellowship.

[About the research presented at the 38th European CF Conference, abstract number ePS06.3:](#)

Neutrophilic airway inflammation is a hallmark of Cystic Fibrosis (CF). Previous studies identified free neutrophil elastase (NE) in bronchoalveolar lavage fluid and sputum as a key risk factor for early bronchiectasis and decline in lung function in CF. Until now, studies have focused on soluble NE activity

in supernatants of airway secretions, while little is known about the relevance of membrane-associated NE activity on the surface of inflammatory cells. In the presented work, NE activity was quantified on sputum neutrophils of 37 patients with CF, using a novel approach based on Foerster resonance energy transfer. Correlations with pulmonary function indices (FEV1 % predicted, residual volume) revealed that membrane-associated NE activity might be a valuable biomarker in CF lung disease.

Rebecca Keyte

Rebecca completed her undergraduate Psychology degree with First Class Honours at Birmingham City University in 2014. Alongside her undergraduate degree, Rebecca worked in a variety of positions within the field of psychology: As a research assistant investigating interpersonal partner coercion and manipulation amongst young people; as a volunteer at “Addactions” drug and alcohol rehabilitation centre; as a volunteer at “Resources for Autism”; and as an “AimHigher” secondary school mentor.



Rebecca was awarded school scholarship and started studying for her PhD at Birmingham City University in 2014. Rebecca’s doctoral research is investigating the role of health beliefs in predicting and explaining risky health behaviours within Cystic Fibrosis patients. Alongside studying for her PhD, Rebecca is a Visiting Lecturer in Psychology.

[About the research presented at the 38th European CF Conference, abstract number ePS03.1:](#)

The increasing life expectancy within the Cystic Fibrosis population is creating manifold challenges for patients in independently maintaining their complex and time consuming treatment regimen. To improve adherence, factors associated with compliance need to be identified.

We therefore investigated the relationships between the beliefs of Cystic Fibrosis adults about their treatment and reported adherence. Participants completed the Hospital Anxiety and Depression Scale and the Beliefs about Medicine Questionnaire. Adherence to pancreatic enzymes, vitamins, physiotherapy and exercise was recorded over two days using daily telephone diaries. It was concluded that in this research, beliefs about treatment did not predict adherence well, demonstrating the need to further investigate the experiences of people with Cystic Fibrosis in relation to health behaviours. Research utilising qualitative interviews is under way to better understand the health related practices and their potential interrelationships with identity and values, and how these relate to adherence. This research will help to develop tools for health professionals to use with young people to discuss risky health behaviours in a way which reflects their experiences and acknowledges the particular issues for this population.



Emma Reece

Emma Reece graduated from University College Dublin with an Honours Degree in Biochemistry and Molecular Biology in 2010. Currently she is a PhD student in the department of Clinical Microbiology in Trinity College Dublin. Her research focuses on *Pseudomonas aeruginosa* and *Aspergillus fumigatus* co-infections in patients with Cystic Fibrosis and understanding how these pathogens interact with each other, the host and how they impact on disease progression. This research is funded by the National Children's Hospital Tallaght.

About the research presented at the 38th European CF Conference, abstract number ePS02.3:

Pseudomonas aeruginosa and *Aspergillus fumigatus* are the most common bacterial and fungal pathogens isolated from Cystic Fibrosis (CF) airways. This research aims to investigate how these two pathogens interact at the CF bronchial epithelial (CFBE) cell surface, contributing to CF lung disease.

P. aeruginosa (mucoid and non-mucoid) and *A. fumigatus* isolates from colonised CF patients were studied. The effect of *P. aeruginosa* and *A. fumigatus* co-infection on the host immune response was examined by ELISAs for pro-inflammatory cytokines, IL-6 and IL-8. The signalling pathways involved in activating this inflammatory response were investigated employing MAP kinase inhibitors and ELISA.

P. aeruginosa isolates inhibited or reduced the growth of *A. fumigatus* isolates in a strain-dependent manner. A greater pro-inflammatory response was observed when CFBEs were infected with the non-mucoid *P. aeruginosa* isolates compared to the mucoid isolates.

CFBE pro-inflammatory response to co-infections was significantly lower than the cumulative inflammatory response predicted. The co-infection induced IL-6 and IL-8 response occurs via the ERK and p38 MAPK pathways.

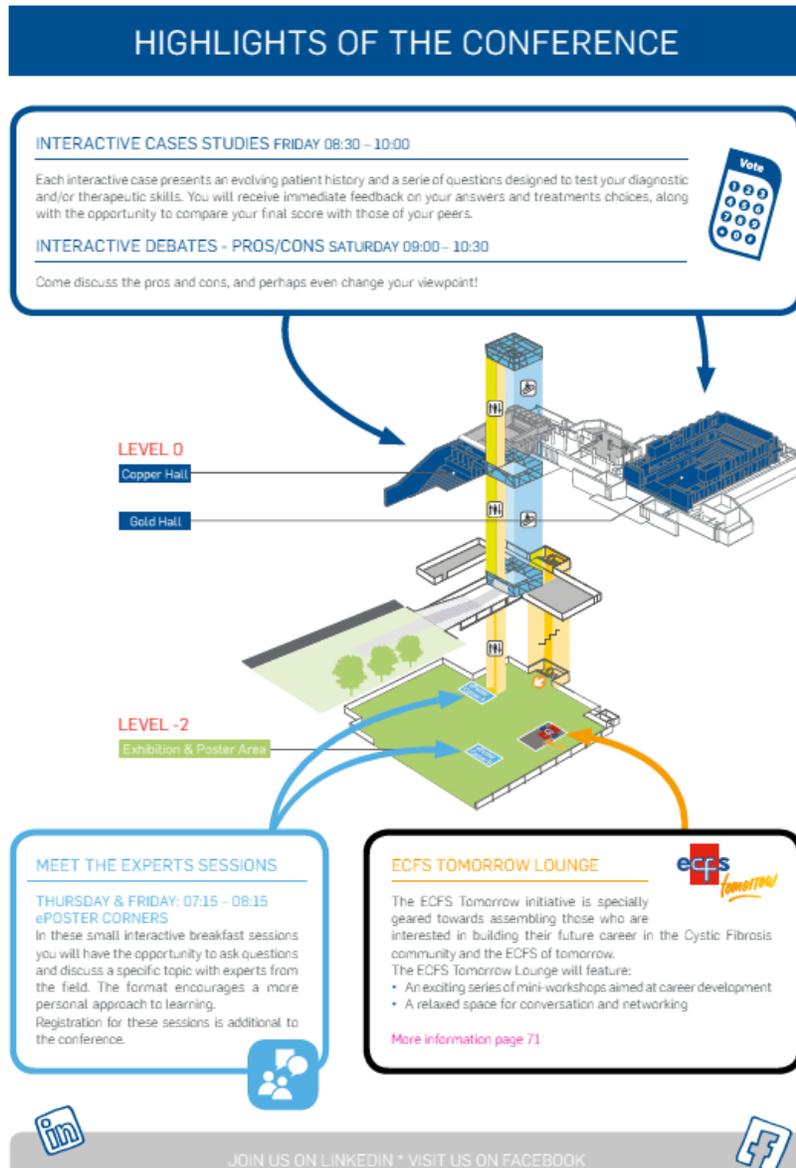
This study demonstrates *P. aeruginosa* is capable of inhibiting the growth of *A. fumigatus*. The competition between these microbes may result in a reduced airway inflammatory response which could allow these microbes to chronically co-colonise the CF airways.

Brussels Conference

The annual ECFS conference taking place in Belgium is fast approaching. My thanks go to the Scientific Committee and the Steering Group for putting together a stimulating programme. This is a long collaboration, which has resulted in the best possible scientific programme and we are excited to provide a forum for the latest news from international experts.

You can find all the details about the scientific programme [here](#).

- Hereafter are some highlights of the conference. **Looking forward to seeing you in Belgium!**



Brussels 2015 Smartphone Application

The number of delegates using the Conference Smartphone App increases every year.

Through the app, you will be able to view information on the scientific programme, the exhibitors, the abstracts and ePosters. The ECFS 2015 Conference App is designed to enhance the experience of the participants before, during and after the Conference. Further information is available on the Brussels website and the App is anticipated ready to download in early June.

ECFS Annual General Meeting @ Brussels 2015

The Annual General Meeting will be held on Wednesday, 10 June 2015, 17:00-18:00, at the Square Conference Centre in Brussels, Hall 400.

The Annual General Meeting agenda:

1. President's Report – Kris De Boeck
2. Secretary's Report – Milan Macek
3. Treasurer's Report – Tacjana Pressler
4. Election results – Milan Macek
5. Update on Journal of Cystic Fibrosis – Scott Bell
6. CTN Report – Tim Lee
7. ECFS Patient Registry Report – Ed McKone
8. ECFS Working Group Reports (click on the hypertext to open the annual reports)
 - [Diagnostic Network Working Group](#) – Nico Derichs
 - [Neonatal Screening Working Group](#) – Kevin Southern
 - [Exercise Working Group](#) – Helge Hebestreit
9. Presentation of the 2016 European CF Conference – Marc Chanson
10. Any Other Business

Meetings at the Brussels Conference

In addition to the scientific programme, there are the following courses and special group meetings

TUESDAY, JUNE 9

08:15 – 16:45	Physiotherapy Short Course – Beyond the lungs – Musculoskeletal dysfunction in CF**	Room 211+212
08:30 – 17:30	ECFS Board Meeting*	Room 202
08:30 – 17:00	Quality Management / Quality improvement Course**	Room 204

WEDNESDAY, JUNE 10

08:00 – 16:30	International Nurse Specialist Group/CF Meeting**	Room 201 A/B
08:15 – 12:30	Physiotherapy Short Course – Beyond the lungs – Musculoskeletal dysfunction in CF**	Room 211+212
08:30 – 15:30	ECFS CTN Training & Development*	Room 202
09:00 – 18:15	CF Course**	Room 204
09:30 – 12:30	CFE/ECFS Joint Symposium - Access to new therapies: How can patient organisations, researchers, health professionals, authorities and industry contribute?	Hall 400
09:00 – 13:45	ECFS Exercise Working Group*	Room 311
09:00 – 16:30	European Cystic Fibrosis Nutrition Group Meeting	Room 213+215
09:30 – 15:00	ECFS Neonatal Screening Working Group Meeting	Arc Room
09:30 – 16:00	European Psychosocial Special Interest Group (EPSIG) Meeting**	Room 206

12:30 – 14:00	International Physiotherapy Group for Cystic Fibrosis (IPG/CF) - Annual General Meeting*	Room 211+212
13:00 – 16:00	Journal of Cystic Fibrosis (JCF) Editorial Board Meeting*	Room 203
13:15 – 17:00	ECFS CTN Steering Group Meeting*	Room 202
14:00 – 16:00	ECFS Patient Registry Executive Committee Meeting*	Room 311
14:30 – 17:00	Physiotherapy Case Presentations	Room 211+212
15:15 – 17:00	ACTIVATE-CF Meeting*	Arc Room
16:00 – 17:00	ECFS Patient Registry Harmonisation group Meeting*	Room 311
16:00 – 18:00	CF Pharmacists Meeting	Room 214-216
17:00 – 18:00	ECFS Annual General Meeting*	Hall 400

THURSDAY, JUNE 11

12:30 – 14:30	ECFS CTN Standardisation Committee Meeting	Hall 400
12:30 – 13:30	ECFS Patient Registry Data Quality Project Group Meeting	Hall 100
13:00 – 14:00	UK Newborn Screening Special Interest Group Meeting*	Silver Hall
13:30 – 14:30	ECFS Patient Registry – Meeting for Interested Parties	Hall 100
14:00 – 16:00	UK Cystic Fibrosis Trust Meeting*	Room 202
18:30 – 20:30	ECFS CTN Blood Inflammatory Markers Standardisation Group Meeting*	Gold Hall
18:00 – 19:30	ECFS Patient Registry Training Reports & Encounters*	Arc Room
19:00 – 20:00	European CF Registry Forum*	Copper Hall

FRIDAY, JUNE 12

12:30 – 14:30	ECFS Diagnostic Network Working Group Meeting	Hall 100
12:30 – 14:30	ECFS Patient Registry Steering Group Meeting*	Silver Hall

SATURDAY, JUNE 13

13:30 – 18:00	ECFS Scientific Committee Meeting - Basel 2016*	Room 213+215
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* closed meeting ** extra registration required

Care-for-Rare Science Award 2015

Many rare diseases remain incurable up to date. Only through enhanced research activities and international cooperation the fortune of many children can be changed. The Care-for-Rare Foundation supports children cross-borders in order to give them hope for treatment – no matter what nationality, ethnic origins and financial background they present.

Endowed with 50.000 Euro, the Care-for-Rare Science Award, sponsored by the Werner Reichenberger Foundation, should give young scientists the chance to initiate a basic or clinical research project in the field of rare diseases. The science award shall promote creative scientific ideas, contribute to a better understanding of the biological mechanisms leading to rare diseases and to the development of new diagnostic and/or therapeutic strategies. Criteria for the selection of the award winner are scientific

excellence, interdisciplinarity and project relevance for clinical-translational research involving the affected patients. The focus is on rare diseases which manifest during childhood.

Application Deadline: 15 August 2015

Please click [here](#) for more details

Basel 2016

Looking forward to 2016, it is a pleasure to announce that Dr. Marc Chanson, Prof. Jürg Barben and Prof. Alex Möller have accepted to respectively be the President and Vice-Presidents of the conference in Basel.

Upcoming Events

- ECFS Board Meeting - 09 June 2015 - Brussels, Belgium
- ECFS Annual General Assembly – 10 June 2015 – Brussels, Belgium
- [38th European CF Conference - 10-13 June 2015 – Brussels, Belgium](#)
- [2015 European Respiratory Society Congress - 26-30 September 2015 - Amsterdam, The Netherlands](#)
- [North American CF Conference - 08-10 October 2015 - Phoenix, AZ, United States](#)
- DNWG meeting – February 2016 – London, United Kingdom
- [39th European CF Conference – 08-11 June 2016, Basel, Switzerland](#)

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