

European Cystic Fibrosis Exacerbation Working Group

Introduction

The CF exacerbation working group was established in June 2018 following an application to the ECFS announced at the close of the 2018 meeting. The objective of this group is to improve the diagnosis, monitoring and treatment of Pulmonary Exacerbation (PEX). Plans for 2020 and 2021 have been impacted significantly by COVID19 both in terms of our inability to meet in person, and clinical staff being redeployed to local respiratory services during the initial peak of the pandemic, as well as the secondary effects of this on local services following lockdown. As the global impact of COVID has continued we have also seen a decrease in exacerbation visits in clinics due to the combined effects of the highly effective CFTR modulator role out and lockdowns. As such the field of exacerbation, inflammation and disease trajectory may have changed for most patients taking modulators.

Group Members

Robert Gray (lead-coordinator), Damian Downey, Cliff Taggart, Don Urquhart, Michal Shteinberg, Isabelle Durieu, Quitterie Reynaud, Silke van Koningsbruggen-Rietschel, Tania Pressler, Malcolm Brodrie, Barry Plant, Stuart Elborn, Ed McKone, Carlo Castellani.

Group Aims

- Re-map CF exacerbations in the post-modulator, post-pandemic era
- Perform systematic review of present definitions of exacerbation as baseline
- Establish the basis for a precision medicine approach to PEX management by creating a platform for the collection and integration of individual patient data (IPD) and patient self-assessment with ECFS registry data
- Establish a programme of work to investigate the utility of biomarkers in the prediction and assessment of treatment response exacerbation, and translate these findings to clinically useful tests
- Utilise our links with the ECFS CTN to develop projects in the areas of biomarker assessment and pulmonary imaging in relation to PEX
- Interact with members of the North American STOP programme to facilitate further exacerbation research and aim to establish a firm basis for a future international collaborative network

Meeting NACF October 2019

A 2hr meeting was held at the NACF where PIs were able to discuss the further plans going forward although it was felt that a meeting where all PIs could attend in early 2020 would be the most productive course of activity as the work streams above were pursued.

Meetings 2020/2021

We have restricted activity to meetings of the groups coordinators to avoid meeting fatigue of members and to allow the pandemic and modulator introduction to come to completion before we can make firm plans for future directions. This has been cost saving and reserved our budget for when we can start facilitating meeting again and planning applications.

Coordinator meetings

Over the pandemic we have had 6 monthly holding strategy group meetings (RG, DD and CT) to develop plans to get the group back to working capacity post-pandemic.

Major Problems at present

- Running a working group during the pandemic to address a specific area of CF management has been challenging. Online meetings do not really afford the same value in terms of idea generation and completion of work
- CF exacerbation has likely changed for many people and we must adapt to address this, whilst considering exacerbation in those unable to take highly effective CFTR modulators

Solutions

- We will hold a half day workshop in early summer to re-address the issues facing on zoom
- Plan a face-to-face meeting late summer to formulate plans to address changing face of exacerbation

Present Outputs

- Systematic review of exacerbation definitions to be submitted summer 2021 (abstract attached)

Robert Gray, Edinburgh, May 2021