

Pulmonary Exacerbation Working Group

Year of report: 2023-2024

Date of initial approval of working group: 2019, renewed 2023

Coordinator: Robert Gray

Long term aims of the working group:

1. To facilitate multi-centre CF pulmonary exacerbation (PEx) research and formulate identification of these events to enable to improve the prediction, diagnosis and monitoring of PEx.
 2. To investigate the utility of biomarkers in the prediction and assessment of treatment response in PEx and develop these towards clinically useful tests.
 3. To explore if a precision medicine approach is feasible by utilising individual and registry patient data as well as assessing the potential of new technologies and biomarkers, such as near patient testing.
- Our approach is outlined in the sections below.

Outcomes already achieved:

In the past year the following outcomes have been achieved by members of the WG:

- Brodlie and Gray scoping review of PEx definition that is currently under review at BMJ open respiratory
- Downey and Taggart publication of paper in ERJ investigating inflammation phenotypes using biomarkers in frequent exacerbators PMID: 38135443
- De Rose and colleagues, European registry data request and analyses of factors associated to more frequent and severe pulmonary exacerbations in CF patients. The data will be presented at the next ECFS Conference in Glasgow.

Report for this year

Summary: The exacerbation WG funding was renewed following application in Autumn 2023. Our central aims remain the same going forward.

Short term goals for the year

1. Continuing work on updated definition of exacerbation. Patient engagement through CF Europe and CF trust are key to this as well as closely liaising with groups active in exacerbation such as STOP. Our proposed Delphi has taken longer than predicted to action and this will be a focus of the next 12 months.
2. Further analysis of European registry data. Dr Virginia De Rose and team are completing the analysis of data related to CFTR modulators effect on chronic airway infections and acute exacerbations. This data will allow hypothesis generation for future studies.
3. Prepare future perspectives in CF exacerbation article. Aim to have article ready to submit within 12 months. This wide-ranging article will include perspective on clinical presentation, patient reported

outcomes, translational science and the impact of CFTR modulators. It will be grounded in the work from aims 1 and 2.

4. Develop new exacerbation studies for submission to funding bodies. This will be led by Dr Stephanie Thee and Dr Quitterie Reynaud, with the support Robert Gray to highlight areas of unmet need in PEx for formulation into potential outline grant applications for the group.

Additional Information

- current number of members 15
- measures taken to encourage ECFS membership: all members of the exacerbation group are expected to be members of the ECFS
- outcomes/achievements (e.g. meetings, activities, website development, awards, publications etc). See section above

Aims for the coming year

- Bi-monthly Zoom meetings of PEx WG
- Business meeting June 2024 at ECF in Glasgow with appointment of new coordinator
- Workshop meeting in October 2024 with focus on data presentations of current projects, possible grant proposals and potential publications as noted in sections above

Summary:

The exacerbation working group had a slow start due to covid and the introduction of modulators. We have now started to produce effective outcome from the group and will move forwards with the aims stated above. A new coordinator will refresh the management of the group and we would suggest that this is assessed annually due to the additional workload for the coordinator above other group members.

Breakdown of expenses (please include total amount received as well as expenditure and, if applicable, the outstanding balance (Euros)): 28K asked for in renewal. So far first spend will be meeting room hire at ECFS Glasgow.

Budget amount requested for next year (please give the amount in Euros and the year): 8K for in person meeting in October

