

## **ECFS PULMONARY EXACERBATION WORKING GROUP**

Please use this template for your annual working group report. Please use black ink, Calibri font size 11. For more information, please read the ECFS working group Terms of Reference document:

Year of report: 2024-2025

Name of Working Group: **Pulmonary Exacerbation**

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

Title, name and email of coordinator, vice coordinator and assistant (if applicable):

Lead Coordinator: Prof. Dr. Virginia De Rose –

Co-Coordinators: Prof. Damian Downey -

Prof. Cliff Taggart -

Dr. Tania Pressler -

Long term aims of the working group (maximum 100 words) including estimated achievement date:

*The long term aims of the PEx WG are:*

-to address unmet needs and knowledge gaps in the field of CF PEx, and advance scientific understanding of these events at a multilevel profile;

-to foster scientific interactions and collaborations across ECFS members and with members of other Societies and to promote and facilitate multicentre clinical/ translational studies.

In particular, the WG will address the following objectives:

- to update and develop a more standardized PE definition;
- to further investigate biomarkers predictive/prognostic and useful to monitor the treatment response in PEx
- to explore the role of digital health and new advanced technologies for early recognition and monitoring of exacerbations
- to review and discuss current treatment and future perspectives in the context of a personalized medicine
- to maintain close cooperation with the ECFS Patient Registry, CTN and other professional groups within the Society

Outcomes already achieved (maximum 100 words):

- A Symposium on Pulmonary exacerbations has been organized at the ECFS Conference in Glasgow
- In person meeting in Glasgow and several online meetings have been held (June 2024- April 2025)
- A scoping review of PEx definitions in CF has been published in *BMJ Open Respir Res* (Brodie, Gray and coll.)
- Analysis of ECFS Patient Registry data: 1) a paper assessing changes in microbiological status of pwCF after ETI treatment has been published in *ERJ Open Res* (Pollak, De Rose and coll.), 2) another manuscript investigating the risk factors of PEx and the effects of ETI on PEx and chronic airway infections will be submitted soon (De Rose, Orenti and coll.). Preliminary results of these studies have been presented at the ECFS Conference in Glasgow.
- A short communication addressing challenging lung infections and clinical trial development in CF has been published in *J Cyst Fibros* (Downey, van-Koningsbruggen-Rietschel and coll.).

Report for this year (max 1000 words)

- short term goals for the year
- current number of members
- measures taken to encourage ECFS membership
- outcomes/achievements (e.g. meetings, activities, website development, awards, publications etc).

- short term goals for the year

- **Update of PE definition:** One of our main goals this year will be to significantly advance the process for developing a new consensus-based definition of pulmonary exacerbation in people with CF. A subgroup and a wider Task Force (including also Registry Representatives and Representatives from CF Europe and the ECFS Physiotherapy SIG) have been set up. Some online meetings have already been held and a plan has been finalized (see below). This will involve a stepwise process where firstly a survey of health professionals and patients is going to be done, secondly an e-delphi process will be held to rate individual components that could potentially be used in a definition(s) and then ultimately to reach consensus on a new definition(s).

- **Submission of an article** on unmet need and future perspectives in CF exacerbation; a draft will be circulated shortly and further discussed during next meetings; the submission is scheduled by the end of the year.

- **Proposals for future research**, including further studies on Registry data. The interest and opportunity of establishing a research network to facilitate new collaborative exacerbation studies at European level and beyond is a major WG long-term goal. There are still many open questions related to exacerbations in CF patients and many aspects to be addressed. There was a preliminary discussion and the WG analysed how to implement this activity as well as the possibility of applying for public/private fundings. Proposals submission was strongly encouraged and will be analysed and discussed during the next in person meeting. In this context, M Shteinberg, a WG member that is also member of the ERS Clinical Research Collaboration "EMBARC", submitted to the WG a project proposal, addressing several aspects of CF and non-CF bronchiectasis that will be evaluated by the WG.

-We are also planning **a series of Webinars** addressing Hot Topics in the field of PEx in order to stimulate the interest and the debate in the field and possibly support the increase of WG membership.

-Two in person **meetings** are planned : the first one will be in Milan during the next ECFS Conference; the second one is scheduled in September/October and will focus on data/results presentations, a further discussion of current projects, an assessment and selection of project proposals and discussion on goals/plans for the coming year. Additional online meetings will also be held.

- current number of members

• Current number of members: 18 (we recently received some additional membership requests that- as far as we know- will be finalized soon).

- measures taken to encourage ECFS membership

-all members of the WG are expected to be members of the ECFS. During the WG meetings it was emphasized the importance of encouraging WG membership, particularly among younger colleagues. Interactions with other International Societies/Groups such as ERS, AMR, and others in which some group members are active can also

provide further opportunities to recruit new members who share the interest in studying exacerbations, encouraging joint membership. A series of Webinars are also planned to stimulate the interest of ECFS community.

- outcomes/achievements (e.g. meetings, activities, website development, awards, publications etc).

At the end of last year there has been a change in WG leadership, as R. Gray resigned and V. De Rose was elected as WG Coordinator. In this context, WG aims and objectives were slightly revised and updated, as well as the WG structure that now includes Representatives from ECFS Patient Registry, CF-Europe and Physiotherapy SIG. This will strengthen and make our interactions and collaboration on common projects more effective. Specific subgroup activities and the plans of action ahead were also revised and further defined during the online meetings.

#### **Meetings:**

-Several meetings have been held by the WG from May 2024 to April 2025 (in person meeting in June at the Glasgow Conference and online meetings in November, March and April) during which we extensively discussed/revised long-term and short-term plans and activities. These included meetings of the subgroup/Task Force on PE definition update with representative from ECFS Patient Registry, CF-Europe and Physiotherapy SIG and the support of Fiona Dunlevy from the ECFS, that will be crucial for setting up the Delphi process.

#### **Activities:**

-*Update of exacerbation definition* : One of our major goal and activity this year was to finalize and launch the process for developing a new consensus-based definition of pulmonary exacerbation in people with CF. A subgroup responsible of advancing this task (Brodlie, Nicholson, Shteinberg, Thee) and a wider Task Force were set up; the process steps were discussed as well as the specific tasks of the subgroup in charge of this activity (primarily of arranging Delphi process). A few online meetings have already been held with all involved colleagues including Registry representatives and representatives from CF Europe and the ECFS Physiotherapy SIG and a detailed plan has been finalized. This will involve a stepwise process which includes firstly a survey of health professionals and patients to ascertain opinions about current definitions, potential differences in exacerbations in the context of highly effective CFTR modulators and to gauge the need for age-specific definitions. Secondly, an e-delphi process will be held to rate individual components that could potentially be used in a definition(s) and then ultimately to reach consensus on a new definition(s). The programme is in progress and the survey is now ready to be launched .

- *A Symposium on Pulmonary exacerbations* was organized at the ECFS Conference in Glasgow.

- *An ERS Research Seminar* “Dissecting the similarities and differences of acute pulmonary exacerbations across chronic airway diseases” will be organized by V. De Rose. The Seminar, recently approved and funded by ERS, will address and debate the impact, complexity and heterogeneity of exacerbations across different chronic airway diseases, including cystic fibrosis, and will foster international collaborative programs focusing on translational and clinical research. WG members will be involved and invited to attend the Seminar and this will be a great opportunity for multicentre, multidisciplinary collaborations and potential novel research projects.

- *Submission of an article* on unmet need and future perspectives in CF exacerbation. An article plan as well as contributions by the interested members and the exact process schedule were discussed (Nicholson in charge of this activity); a draft will be circulated shortly and further discussed during the next meetings; the submission is planned by the end of the year.

#### **- Publications:**

-Scoping review of PEx definitions in CF published in *BMJ Open Respir Res* (Brodie, Gray and coll.)  
 - Analysis of ECFS Patient Registry data: 1) a paper assessing changes in microbiological status of pwCF after ETI treatment published in *ERJ Open Res* (Pollak, De Rose and coll.), 2) another manuscript investigating risk factors of PEx and the effects of ETI on PEx and chronic airway infections will be submitted soon (De Rose, Orenti and coll.). Preliminary results of these studies have been presented at the ECFS Conference in Glasgow.  
 -Short communication on challenging lung infections and clinical trial development in CF published in *J Cyst Fibros* (Downey, van-Koningsbruggen-Rietschel and coll.).

Aims for the coming year (please state year) (max 50 words):

Year: 2025-2026

- To advance ongoing projects, complete our programme of PE definition update and publish the results.
- To start new multicentre, multidisciplinary research projects in the context of our long term aims and in collaboration with other ECFS WG/International Societies
- To continue encouraging WG and ECFS membership
- To develop a Webinars series focusing on Hot Topics in PEx
- Online meetings will be held every two months; a business meeting will be held at the next ECFS Conference in Milan and then two next meetings (workshops) with focus on results presentations and discussion of ongoing projects/activities

Summary (maximum 100 words):

At the end of last year there has been a change in WG leadership , as R. Gray resigned and V. De Rose was elected as WG Coordinator. In this context, WG aims and objectives were slightly revised and updated. The WG aims to address unmet needs and knowledge gaps in the field of CF PEx and to advance scientific understanding of these events. Several projects and activities are now progressing (some of which will be completed this year) and the WG is actively working to provide an infrastructure for facilitating novel clinical/ translational studies at European level and beyond. To reach this goal, the close cooperation with other ECFS Group will remain crucial as well as all the activities to inspire and attract new members.

Breakdown of expenses (please include total amount received as well as expenditure and, if applicable, the outstanding balance (Euros)):

Overview of the PEx WG budget:

Pulmonary Exacerbations Working Group		
<b>Pulmonary Exacerbations WG - 2024</b>	<b>EUR</b>	
	10.000	Opening balance
Income	10.000	
Expenses Pulm Ex meeting GLA	- 46	Pulm Ex mtg GLA
	<u>19.954</u>	

We would like to ask if we could carry-over 15.000 Euro for one additional year; these funds will be a very relevant support for organization of next in person meetings (the first one is scheduled for September/October 2025), administrative/professional support for ongoing projects, in particular for the survey and the delphi process on exacerbation definition update (translating documents to various languages and dissemination, data collection and statistical analysis), publication fees and travel awards for younger members.

Budget amount requested for next year (please give the amount in Euros and the year):

We would like to request the full amount of 10.000 Euro.

**The deadline for submission is 1st May 2025.** Please email the report **and the excel spreadsheet of your members** to: [jess.matthews@ecfs.eu](mailto:jess.matthews@ecfs.eu)

The report will then be presented to the board prior to the summer ECFS board meeting and then a decision will be made at the next board meeting for approval before being distributed.