Meeting of the ECFS-DNWG

NACFC Baltimore, October 22nd 2010

Participants: Yasmin Yaacov, Hugo De Jonge, Inez Bronsveld, Kris De Boeck, Ernst Rietschel, Silke Van Koningsbruggen, Nico Derichs, Anne Munck, Isabelle Fajac, Harry Cuppens, Kevin Southern, John Dodge, E Gaillard, Marisa Sousa, Paola Melotti, Lisa Kent, Els van der Wiel, Lutz Naehrlich, Gilles Rault, Luisa Pereira, Celeste Barreto.

Kris De Boeck introduces the different participants of the DNWG that will give an update on present matters of the DNWG.

Document outcome parameters from the ECFS-Clinical Trials Network (following meeting in Venice). Kris and **Lisa Kent** already drafted the introduction (definitions of outcome parameters and clinimetrics) and started (re)drafting the clinimetrics tables and the answers to the questions. The working groups on 6 types of outcome parameters (for use in clinical trials, not for diagnostics!) delivered a lot of information. This info is now being presented in a very uniform and orderly manner. Lisa and Kris will send the (re)drafted tables back to the different working groups for review. One representative per working group needs to check whether a full literature search was indeed performed. In addition we agree on the interpretation: e.g. positive evidence means 3 or more conclusive publications on a subject; negative means 3 or more studies with negative evidence; less than 3 studies or contradictory data are reported as insufficient evidence.
To brief each working group well 2 teleconferences per group are planned with Kris, Lisa and the group representative(s). The first to clearly explain the global plan and the document. The second to hear the comments and suggestions from the group.
The biomarker group provided insufficient info about sweat test. Rather than omitting it there will be a catch-up: Marijke Proesmans (Leuven) and Nguyen (Paris) will do the work. This will then be reviewed by Kevin Southern and other initial sweat test co-workers.
Time line for this report: before end of the year.

Next we discussed new issues for the program in Stockholm, Friday and Saturday February 11th - 12th, 2011.
Please adjust your travel plans to fly in on Thursday February 10th and fly back on Saturday February 12th afternoon.
Suggestions for the program:

1. CFTR2 project clinical data: Garry Gutting, John Hopkins
2. Terminology in CF diagnoses: Delphi consensus: send it out by the DNWG with help of the ECFS. We need to make sure that a sufficient number of adult pulmonologist are included in the survey. Option can be to send it via ECFS
3. Diagnositic tools: monocytes (update Paola) pancreas associated protein: IRT-PAP versus IRT-DNA protocol: Hugo de Jonge/Jeanette Dankert
4. NPD ratio's including results combined with other outcome parameters. MW
5. ICM: update SOP multicenter comparison between reference centres. ND
6. Skin conductance. Fajac
7. Tools for interpreting consequences of mutations
8. the CFTR2 project: in vitro data
9. Swedish contributions: X-ray/NPD solutions
10. ECFS website: uploading files and using it as an interface to discuss cases.
11. Interesting patient cases: time permitting
12. Do we want to become an official ECFS working group? Pro and con's

13. As in former years four young investigators can participate in the meeting with a grant of 500 euro's. Contributions can be e-mailed to Michael Wilschanski.

Nico Derichs reported on the progress of the ECFS ICM SOP:
-ICM protocol (de Jonge 2004) now validated for difficult CF diagnosis (Derichs 2010)
-European ICM survey (6 centres) confirms need of standardisation, to be published
-ECFS ICM SOP: joint SOP DNWG (CF diagnosis) & CTN (outcome marker), final version approved, all main details harmonised with CFF TDN ICM SOP draft, reference/training centres in place (Rotterdam/Utrecht, Berlin), multicenter comparison study (Rotterdam/Utrecht, Berlin, Verona, Jerusalem?) will start in November 2010.

Inez Bronsveld showed the last details of the NPD SOP: the last version has been sent to the NPD working group participants.The few issues that are being solved now are:

- 1 uniform catheter by Marquat company
- Preparation of perfusion solutions without precipitation
- Sequence of the measurements, do we need to measure AT and centimetre tracked measurements.

Last comments can be e-mailed to I.Bronsveld@umcutrecht.nl. SOP will be finalised October / November 2010.

To become a member of the ECFS (which includes a membership to the Journal of CF and discount for the ECFS meeting) please contact Christine.dubois@ecfs.eu

Inez Bronsveld, Kris De Boeck