

Group meeting ECFS Diagnostic Network Working Group (DNWG), at ECFC in Valencia on Friday June 18, 2010, at 13:00-15:00

Final results of the "Warming of the Nasal Potential Difference (NPD) solutions" study

The DNWG has been discussing whether warming the NPD solutions to 37°C has an effect on chloride transport. Inez Bronsveld presented the results of the warming studies: NPD measurements were performed with solutions at room temperature which were then changed to solutions at 37°C. CF centres that performed these studies were: Leuven, Brussels, Jerusalem, Verona, Sydney, and Utrecht.

In the analysis of the whole group it did not appear to make a significant difference between warming the solutions or using room temperature solutions. However, a subanalysis performed by Leuven showed a significant difference between warm and room temperature solutions of about 2 mV. Statisticians from Utrecht and Leuven will be collaborating to evaluate the whole data set again.

We observed that in all participating centres the variability of the measurement increased with warmed solutions: SD and range of the responses increased. Because of this increased variability, although there seems to be a slight difference between warmed and room temperature solutions, we voted in our DNWG that we will not include warming of the perfusion solutions in our standard operating procedures (SOP). For study purposes we will use protocols demanded for by the sponsor. However, when sponsors contact us about a study with NPD measurements included, we will discuss our own SOP with them, including the use of room temperature solutions.

The Delphi Consensus for the clinical diagnosis of CF

The DNWG is working on a Delphi consensus for the clinical diagnosis of CF. Kevin Southern reported on the progress of this Delphi consensus. It is the idea to come to a consensus for the clinical diagnosis for CF. In Paris we voted for an 80% agreement for this consensus.

Different co-workers are now preparing the appendices of the document for different outcome parameters, like clinical features, sweat test, *CFTR* gene analysis, NPD / Intestinal current measurements (ICM).

When these appendices are finished the document will be send out to all the members of the DNWG to be filled in.

Planning New Studies in the DNWG

1. After standardisation of the different NPD protocols used, we will evaluate whether this standardisation has influenced the variability in results. We will analyse whether our standardisation has decreased variability within one centre and between the different centres.

2. The plan is raised to create a database representing all atypical CF cases, with inclusion of as much parameters as possible like: clinical features, *CFTR* mutation analysis, sweat test, and electrophysiology.

DNWG Website update

Inez Bronsveld explained the latest developments on our website facilities. All ECFS members have a website profile. When you log in on the ECFS website, DNWG members will see a DNWG directory. We can file documents in this directory, guidelines, protocols, and for instance exchange difficult NPD and ICM tracings for interpretation. Patient data have to be used anonymously.

We concluded that it is fair that you have to be an ECFS member, since the ECFS is facilitating this website and also aids us with the development towards new features.

DNWG Stockholm 2011 meeting

Lena Hjelte announced the next DNWG meeting in Stockholm on February 11 and 12, 2011. Just as last year there will be a competition for Young Investigators to attend and receive a travel grant. Young Investigators can send their abstract to Michael Wilschanski, e-mail: michaelwil4@gmail.com; deadline December 31st, 2010.

Report by Michael Wilschanski and Inez Bronsveld