

# Minutes digital IPSCSG biannual meeting

Date: 30 Oct 2020

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## Agenda:

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|--|-----------------|
| 1. Minutes meeting Boston November 2019                                  | Cyriel          |
| 2. Draft programme 6 <sup>th</sup> biennial meeting June 2020 London     | Doug/Steve      |
| 3. News and proposals from the Working Groups:                           |                 |
| ○ <i>MRI</i>   |                 |
| ▪ upcoming MRI online meeting November 2 <sup>nd</sup>                   | Christoph       |
| ○ <i>CCA</i>   | Luca            |
| ▪ novel murine PSC-CCA model   | Yuri Popov      |
| ○ <i>Natural History, Biomarkers, Clinical Trial Design</i>              |                 |
| ▪ FICUS  | Olivier/Cyriel  |
| ▪ R-Liver  | Christoph       |
| ▪ proposal: tofacitinib in PSC case series                               | Christoph       |
| ▪ IPSCR  | Cyriel          |
| ▪ CALiD  | Cynthia         |
| ▪ Retrospective DB: current projects                                     | Tobias          |
| ▪ Ancient genomes in PSC   | Alessio Gerussi |
| ▪ proposal: retrospective study on endoscopic management of advanced PSC | Joshi Deepak    |
| ▪ proposal: pregnancy and PSC  | Shanika Nayagam |
| ○ <i>Transplantation</i>   |                 |
| ▪ recurrent PSC project  | Palak           |
| 4. Rotations of SC members and WG leaders                                | Ulrich          |
| 5. Other   | Cyriel          |

## Ad 1.

The minutes of the November 2019 meeting in Boston were arrested.

## Ad2.

Doug mentions that the proposed time for the postponed Biennial meeting in London is September 2021. Funding is secured. Care has to be taken there will be no conflict with the UK Hepatology meeting. PSC Support and potentially PSC Patients Europe have expressed their wish to organize a meeting in conjunction with the IPSCSG Biennial meeting. This is welcomed.

Josh Korzenik mentions that PSC Partners for Cure have also expressed their interest in a meeting back to back. This may have to be organized through digital means then.

## Ad 3.

### *MRI WG:*

Christoph announces the next MRI WG Zoom meeting, which will take place in three days on November 2<sup>nd</sup>. The invitation has been circulated several times.

*CCA WG:*

Luca alludes on the European CCA registry, which now contains almost 3600 patients from 30 institutions. Furthermore, he mentions the project “Novel protein biomarkers in serum extracellular vesicles for the diagnosis of cholangiocarcinoma (CCA) in patients with primary sclerosing cholangitis (PSC)” The validation phase is now completed and a manuscript in preparation. Another project, Metabolomic-based prediction of prognosis in patients with PSC and early diagnosis of cholangiocarcinoma: new non-invasive strategy is led by Jesus Banales and has now entered its validation phase. More cases, from which 300 microliter of serum is required are welcomed.

**Action point: Luca will send Cyriel a template what is needed exactly to be circulated among the IPSCSG and listed on the website.**

Lastly, Luca presents a new PSC-CCA animal model, a MDR-/- thioacetamide induced mouse model.

Yuri Popov extensively describes another novel PSC-CCA mouse model using Sleeping Beauty transposon-transposase system (SB Md2-/- .CCA model).

Luca and Yuri are open for a liaison in order to cross-pollinate their experiences.

*Natural History, Biomarkers, and Clinical Trial Design WG:*

Cyriel presents on behalf of Olivier the progress of the FICUS study. The study data collection will be completed in June 2021. Participants are encouraged to keep filing data in this important project. Upon completion of FU and biobanking a steering committee will decide which projects will be conducted with the precious biomaterial that has been collected in the past 5 years.

Christoph describes R-LIVER, which is a prospective multicenter registry on incident cases with rare liver diseases across many European countries, including paediatric, transition and adult patients. It uses Castor EDC as a platform and complies with the definitions of data elements as agreed upon by the IPSCSG in 2016. The registry plans to develop an App for PRO capture.

Christoph presents a proposal to study the response of the liver in PSC/IBD patients who use tofacitinib. Cyriel asks whether the three months exposure to tofa is mandatory. This may not be necessary.

**Action point: please contact Ida Schregel or Christoph ([i.schregel@uke.de](mailto:i.schregel@uke.de) or [c.schramm@uke.de](mailto:c.schramm@uke.de)) if you have any patients who are/have been using tofacitinib. Ida/Christoph will produce a full protocol and template what data are needed. This will be circulated and put on the website.**

Cyriel presents the current status of the IPSCR. In the Netherlands 919 patients have been entered with their retrospective data and 750 are being followed prospectively. Some 400 will follow soon. In Oslo 37 patients have been entered and 36 in Stockholm. Helsinki is almost ready to import their DB of 944 PSC patients in their new Castor IPSCR DB. A consortium agreement and generic ICF is now being completed.

Cynthia describes the progress with CALiD. Some 972 patients have been entered in the Redcap DB. On 162 cases there now is one year FU. She announces the ItCH-PSC study, which will study pruritus in PSC in 6 centers and will start in Q1 2021.

Tobias gives an update on the studies that are run with the large IPSCSG retrospective DB.

Three studies have been successfully completed and will be published shortly. Another 9 studies are currently running but inclusion is very low. He calls upon the PIs to revive inclusion through advertising.

**Action point: put studies and main inclusion criteria on the website. Each PI is encouraged to promote their studies through the IPSCSG network. Send email to Hadassa at [ipscsg@amc.uva.nl](mailto:ipscsg@amc.uva.nl)**

Alessio Gerussi presents an update on the Archaic DNA & PSC study. Current analyses are exploring whether there is a signal of enrichment in Neanderthal regions/alleles for candidate genes across several immune-mediated diseases. Next step: single phenotypes (including PSC) by using individual data.

Joshi Deepak from King's college London presents a proposal for a retrospective study to look into the benefit of endoscopic treatment of dominant strictures in PSC patients with advanced disease.

Cyriel mentions that a model which is now being developed in the Mayo Clinic in Rochester is answering a slightly different question. He proposes to contact Christian Rupp who is finishing a study in which he has collected data on 1174 PSC patients with dominant strictures.

**Acion point: Joshi will contact Christian Rupp. If renewed data collection is still needed he will circulate a protocol through the IPSCSG, which will also be put on the website.**

Shanika Nayagam from King's college London presents a proposal to study maternal outcome of pregnancy in PSC. Tobias offers to help with choosing centers. The retrospective DB however does not contain information on pregnancy.

**Action point: Shanika will write a proposal and have this circulated through the IPSCSG network including listing on the website.**

Palak presents an overview of the multi-centre DB on PSC-patients listed for transplant. Aliya has made a Redcap based DB to supplant the previous DB.

There are currently 7 research proposals.

**Action points: Palak and Aliya wil circulate through IPSCSG network the inclusion/exclusion criteria for patients and will put the proposals on the website.**

#### Ad 4.

Ulrich presents a proposal from the secretaries and supported by the Steering Committee to postpone rotation and new candidacy of Steering Committee members and working group leaders until the next Biennial Meeting in September 2021. No one objects.

#### Ad 5.

There are no other items or remarks.

Cyriel thanks everybody for their attendance and active participation and PhD fellow Bregje Mol for her assistance to run the Teams meeting smoothly,

The next meeting will hopefully be in person during ILC2021 in June 2021 in Amsterdam.