Meeting minutes ESPN Working Group Glomerular Diseases
54th Annual Meeting, Ljubljana, Slovenia
23-06-2022; 7:30-9:00

1. Welcome

2. Upcoming elections – board members
   - Rezan Topaloglu will resign as board member of the GD working group. All members are invited to apply for board member, which is a position for 3 years.
   - List of publications from the working group (not discussed in detail due to time restrictions)

3. Update guideline projects (by Olivia Boyer)
   - Consensus statement on CNS has been published, free webinars are available online.
   - Practice recommendations SRNS, including patient documents are published.
   - SSNS practice guideline, the core group is led by Dieter Haffner. Meetings (1-2/week) were held online, which made it rather challenging.
   - Extended review of evidence, however, evidence was limited. Still, recommendations have been made. The manuscript has been submitted.
   - Definitions:
     - Adjusted: FRNS: reduced to ≥3 relapses/year,
     - Introduced: sustained remission, SSNS control on therapy, SSNS not controlled on therapy, complicated relapse.
   - Indications for kidney biopsy:
     - A biopsy is not indicated at first onset, only in children with atypical features of INS or in populations with high risk for IgA. In case of infantile NS (age 3-12 months), biopsy in combination with next generation sequencing is indicated.
     - Biopsies should never be performed before start of CNIs, only if exposure has been more than 2 years.
   - Steroid therapy:
     - An 8-week schedule is encouraged: 4 week 60 mg/m^2/day (max 60 mg/day), followed by 4 week 40 mg/m^2/AD (max 40 mg/day), as a single dose in the morning.
     - No tapering during alternate dosing.
   - Treatment of relapse:
     - Induce remission with 60 mg/$m^{2/}$day (max 60 mg/day), followed by 4 weeks 40 mg/m^2/AD (max 40 mg/dose) after 3 days of remission. No tapering
     - There is no evidence for prednisolone for the prevention of relapses during respiratory tract infection, but escalation of prednisolone therapy (from alternate day to daily) can be considered.
   - Maintenance therapy:
     - The authors found that there is great variation between countries.

    - Steroid-sparing agents are indicated in FRNS/SDNS. Shared-decision making (balancing desired and adverse effects) with parents is highly recommended.
    - Rituximab is recommended as a second-line treatment, when treatment of at least 1 steroid-sparing agent has failed.
- Supportive treatment:
    - No changes.
- Ongoing development of guidelines concerning IgAN and IgAV were shortly mentioned.

4. ESPN evaluation of immune and vaccine competence (by Marina Vivarelli)
    - Inclusion has almost completed, already a large cohort.
    - Baseline (at onset in nephrotic state) results: all patient had significantly lower IgG levels than age- and sex-matched healthy controls. Significant reduction in anti-tetanus/HBV titers. B cell memory cells can be used to evaluate immune status.
        - Q: what happens when patients are in remission? A: IgG levels tend to go back to normal.
        - Q: Advise on vaccination? A: as many as possible when the immunosuppresive dose is at its lowest, but be reluctant with live-attentuated vaccination under prednisolone treatment.

5. Update membranous nephropathy (by Julien Hogan)
    - This presentation was a short update of the one held the previous year.
    - In the participating centres, there was an Increase in PLA2R-screening, both the possibility to and practice of screening increased.
    - The study consists of multiple steps. In Step 3 of the study, unstained slices can be used for discovery analysis.
        - For this, the team applied for an ESPN grant.
    - For potential centres that want to participate to the study: in case IRB issues arise, contact the study team as they can help with documentation.

6. Lupus registry (by Adriana Suhlrie and Rezan Topaloglu)
    - This was an online presentation, hampered by an initially bad connection that gradually improved during presentation.
    - A new ERKNet registry and its inclusion criteria (extended to obtain long-term data) and study design has been presented. The registry has been online since April 2021. Since then, a total of 52 patients have included.
        - Q: Will patients who are entered into the subregistry be automatically entered into the main registry? A: Yes.
    - The registry has been improved by implementing data validation, thereby decreasing the risk of wrongful data entry. Also, parameters at diagnosis were added.
    - All members are invited to add patients to the registry, contact information is presented on the slide.
7. Nephrovir-3 & LEARNS-2 (by Claire Dossier)
    - Introduction to rationale and design of study.

- Between September, 2017 and February, 2020, 86 patients were included in the study of which 63 were randomised. Inclusion was stopped since the data will be pooled with data from the LEARNS study (data sharing agreement finalized).
- Results: there was a significant decrease in risk for relapse at 1 year of follow-up.
    - Q: What was the risk of neutropaenia? A: There was only 1 case of transient neutropaenia, but levamisole did not need to be discontinued.
    - Q: There are problems with the availability of levamisole in Italy, is this more widespread? A: There may be delivery problems, as France and the Netherlands experience similar availability problems. This calls for more European regulation and cooperation.
- Future plans:
    - Data pooling with LEARNS. An individual patient data meta-analysis will be performed.
    - Follow-up of the Nephrovir-3 will be extended to 24 months.
    - LEARNS-2

8. RTX and hypogammaglobulinaemia (by Aleksandra Zurowska)
    - Publication submitted to NDT.

9. Covid-19 and ANCA-vasculitis and Takayasu arteritis (by Matko Marlais)
    - Covid-19
        - Key outcome: severe clinical outcome.
        - There were differences between patients with and without RRT and at last known follow-up (high serum Cr at presentation was associated with more likely receiving RRT).
    - Takayashu arteritis:
        - Data is coming in, the aim is to characterize Takayashu arteritis in children.

10. New ideas and proposals (Kjell Tulus)
    - RTX dosing in FRNS/SDNS:
        - Dose doesn't really matter
        - Repeated doses: there is an increase in efficacy after course 4, without increase in side effects.
    - RTX in SRNS: 23% remission after 6 months.
    - CNI therapy in mendelian SRNS: response in 22.5% of the children, with improved kidney survival.
    - Lupus nephritis: >400 children included in the study. Depending on the definition of remission (7 definitions were used), there was a wide range in achieving remission at 24 months of follow-up (approx. 45-80%). A second paper is currently under review, and 3-4 more papers are coming up.

11. Closing