

Paediatric Vasculitis Working Party

Chair: Marija Jelusic (marija.jelusic.drazic@gmail.com)

Secretary: Despina Eleftheriou (d.eleftheriou@ucl.ac.uk)

To all Vasculitis Working Party members

Dear Colleague,

Here is a short report of Vasculitis Working Party meeting that we had on September 5, from 16:00 to 17:30 during PREs 2018 congress in Lisbon

1. Update from PREs Working Party Strategy meeting with the Executive Board - PREs Working Parties 2.0 (ppt attached)

All WP Chairs met together in London, June 22nd with the PREs President, Berent Prakken, and members of the PREs Executive / Council **to revisit the roles and activities** of the PREs WPs. The **PREs Working Parties 2.0** is launched.

All **PREs Working Parties 2.0** will be characterised by:

- Committing to contributing to and advancing the goals of the **PREs Mission** “Dedicated to advance the care and improve the health and wellbeing of children and young people with rheumatic conditions”
- **Focussing on each of the three ‘PREs pillars’**, namely fostering excellence in 1. Clinical Care, 2. Science and Research, 3. Training and Education
- Having specific defined goals, with aims, objectives and timelines, aligned to each of the three pillars
- Proactively engaging with EMERGE in a demonstrable way
- Having clear terms of roles for the Chair and Secretary, with leads for each of the three pillars

2. Development and Validation of Juvenile VAsculitis Multidimensional Assessment Report (JVAMAR) (update)

The aim of this study was to develop and validate multidimensional assessment instrument, called the “Juvenile Vasculitis Multidimensional Assessment Report” (JVAMAR), to measure all the domains of vasculitis in standard clinical care.

Materials and methods: The JVAMAR includes 12 parent or patient-centered measures and 5 dimensions that assess functional status, pain, disease activity, therapeutic compliance and health-related quality of life (physical, social, school, emotional status) with disease outcome. It is proposed for use as both a proxy-report and a patient self-report. The Outcome Measures in Rheumatology Clinical Trials (OMERACT) filter was applied.

Results: Data set was collected 2013 to 2016 from seven pediatric rheumatology centers in Turkey. The forms were completed by 164 children with vasculitis in 429 visits. Completion and scoring appeared to be quick, requiring < 15 minutes.

The JVAMAR was found to be **feasible and to possess face, content, criterion and construct validity.**

Conclusion: The development of the JVAMAR introduces a new and multi-dimensional approach in pediatric rheumatology practice for the Vasculitis. It is a valid tool for children with vasculitis and will help enhance the quality of care in this group of patients.

For further information please contact: PI – Erkan Demirkaya (dottore_erkand@yahoo.com)

3. Paediatric Vasculitis Damage Index (PVDI) (update)

- plan to do a few final changes to the original draft of paper and submit it to Pediatric Rheumatology Online Journal at the end of the year.

For further information please contact: PI - Pavla Dolezalova (Pavla.Dolezalova@vfn.cz)

2. MYPAN (update)

"An open label randomised controlled trial of mycophenolate mofetil versus cyclophosphamide for the induction of remission of childhood polyarteritis nodosa"

- *Hypothesis:* MMF (experimental therapy) is not inferior to CYC (standard therapy) for induction of remission of PAN within 6 months
- *Primary endpoint:* remission within 6 months (PVAS 0/63 on 2 consecutive readings at least one month apart on protocol steroid taper)

PROGRESS and RECRUITMENT

- **Recruitment closed on 30.06.2018.**
- 11 patients recruited: 7 UK, 4 non-UK

GOSH London - 3; Alder Hey Liverpool - 3; Manchester - 1; Barcelona -1; Ankara - 3

- **Trial end date: 30/04/2020**

For further information please contact: PI - Paul Brogan (p.brogan@ucl.ac.uk)

NEW CLINICAL TRIALS:

4. BIOVAS - "A pragmatic, randomised, double-blind, placebo-controlled, modified-crossover trial of biologic therapy for refractory vasculitis in adults and children".

Now funded by NIHR. Protocol in development.

For further information please contact: PI - Paul Brogan (p.brogan@ucl.ac.uk)

5. KD-CAP (Kawasaki Disease Coronary aneurysm prevention)

"Multi-centre, prospective, randomised, open-label, blinded end-point study of prednisolone plus intravenous immunoglobulin (IVIG) for prevention of coronary artery complications in acute Kawasaki disease"

Funding application submitted to BHF

European sites (considering application to C4C)

Please contact : d.elftheriou@ucl.ac.uk

PROPOSAL FOR PReS WIDE PROJECT

6. Histological classification for predicting outcomes in HSP

Primary aims of the study:

- To validate the feasibility of the modified semiquantitative classification (SQC), to validate the feasibility of the Oxford classification, to validate the feasibility of the Haas classification,
- To compare the ISKDC, Haas, Oxford and modified semiquantitative classification (SQC) for their ability to predict the clinical outcomes in HSP

- To establish which variables of each histological classification most significantly predict renal outcome/have the strongest association with unfavorable outcome

Patients and methods:

- A multicenter (including pediatric rheumatologists - effort of PReS, PRINTO, EULAR? and paediatric nephrologists) combined retrospective and prospective study
- patients aged < 18 years at the time of diagnosis with biopsy-proven HSPN (patients' medical records and appropriate histology material for re-analyses + newly diagnosed pediatric patients)
- biopsy material – reviewed and/or scored by according to ISKDC, Oxford, Haas and SQC classification by an experienced renal pathologist, blinded to the patients' medical histories and laboratory results.

For further information please contact: Marija Jelusic (marija.jelusic.drazic@gmail.com)

7. Announce the IPNA (International Pediatric Nephrology Association) Teaching Course and 9th SEPNWG (South Eastern European Pediatric Nephrology Working Group) Meeting, endorsed by ESPN (European Society for Paediatric nephrology) and ISN (International Society of Nephrology)
- **Main topic "Vasculitis", November 2019, Croatia**

Sincerely yours

Marija Jelusic, Chair of Vasculitis WP

Despina Eleftheriou, Secretary of the Vasculitis WP