

Paediatric Vasculitis Working Party

Chair: Marija Jelusic

Secretary: Despina Eleftheriou

Lead of education and training: Teresa Giani

Lead of clinical care: Neil Martin

Lead of science and research: Ezgi Deniz Batu

EMERGE representative: Mario Sestan

To all Vasculitis Working Party members

Dear Colleague,

Here is a short report of the Vasculitis Working Party meeting that we had on September 25th, from 11:00 to 12:30, during PREs 2020 e-Congress.

1. ACTIVITIES FOR CLINICAL CARE AND RESEARCH

- **George Reynolds "Introduction to the Vasculitis Federated & Model registry"** (ppt attached)

Mr. Reynolds has worked as a consultant and policy advisor on rare disease registries since 2013. For the last year he has been developing a *pan-European approach to vasculitis patient registries for EUVAS the European Vasculitis Society*. This is based on a core common registry; the Model Registry using the REDCap clinical data collection platform. This model registry is available free of charge to all researchers. In advance of its release, the registry has been extensively reviewed and updated by the EUVAS registry and annual assessment working group. Ten countries have expressed an intent to implementing the model registry. Several countries with an existing funded vasculitis registry have committed to supporting the annual assessment minimum data set. In the long term this will allow patient treatment and outcome data to be shared across Europe. This approach is based on the European Cystic Fibrosis Registry that Mr. Reynolds advised for several years.

For more details please contact: George Reynolds (george.reynolds@rareurn.eu)

- **Pavla Dolezalova "Collaboration with ERN-RITA" and "Introduction to EUVAS"**

Paediatric vasculitis within paediatric rheum disease stream, vasculitis in general is the main group of the autoimmune stream (chaired by Mark Little)

Educational activities: Regular webinars or RITA diseases provided by RITA experts incl vasculitis, please check RITA website <https://ern-rita.org>

Common projects

EUVAS registry – for further discussion

- Proposal: Pavla will invite a core group to meet with Mark Little to formulate a proposal for the PREs vasculitis WP community regarding options for paediatric vasculitis data collection

- Update of the SHARE vasculitis guidelines to be discussed also in collaboration with the RITA guidelines working group (chaired by Andrew Genery)

2. EDUCATION AND TRAINING ACTIVITIES

- **Rolando Cimaz** - announced the **First European Congress on Kawasaki Disease EURO – KiDs Congress**, from 14 to 16 January 2021, at the CIS Auditorium - Institut Pasteur, Paris, France

A first event in Europe! A half-day workshop: "Kawasaki disease, autoinflammation and beyond" aimed to gather experiences on new treatment targets, biomarkers, and integrated research strategies.
A two-day main meeting aimed to update on clinical innovation, recommendations, basic science, and sharing clinical experiences.

For more information please contact: Rolando Cimaz (rolando.cimaz@unimi.it)

- **Teresa Giani** – announced the **1st PReS Educational Course "Vasculitis in paediatric age"**

WHEN: 2021, HOW: hybrid meeting / F2F, WHAT: 3-day course, comprehensive training on main systemic vasculitides, new multidisciplinary diagnostic approaches, latest scientific advances, recent recommendations and guidelines for the treatment. *The registration will be open to all PReS members and will be free of charge.* Waiting for the final approval of the PReS.

For more information please contact: Teresa Giani (teresa.giani@gmail.com)

3. SCIENCE AND RESEARCH ACTIVITIES

3A. Marija Jelusic: "Histological predictors of outcome in patients with Henoch-Schonlein purpura / IgA vasculitis and nephritis"

Primary aims of the study: to compare the four most commonly used histologic classifications (ISKDC, Haas, Oxford and SQC) for HSP/IgAV nephritis, to determinate which one is the best predictor of disease outcome and to establish which variables of each histological classification have the strongest association with unfavorable outcomes

Patients and methods: A multicenter study, patients aged < 18 years at the time of diagnosis with biopsy-proven HSPN/IgAVN (patients' medical records and appropriate histology slides for re-analyses)

PROGRESS: Abstract: "DIFFERENT HISTOLOGICAL CLASSIFICATIONS FOR HENOCH-SCHÖNLEIN PURPURA NEPHRITIS - WHICH ONE IS THE BEST PREDICTOR OF DISEASE OUTCOME? PILOT STUDY OF THE PReS VASCULITIS WORKING PARTY." M. Held, M. Sestan, M. Coric, S. Bulimbasic, T. Giani, N. Martin, N. Kifer, S. Srsen, D. Kifer, M. Heshin, A. Ravelli, R. Cimaz, S. Ozen, A. Gagro, M. Frkovic, M. Jelusic on behalf of the PReS Vasculitis Working Party - has been accepted for a Lightning talk at the PReS 2020 (presented by our EMERGE representative Mario Sestan) and as a poster at the ACR Convergence 2020

We would like to invite all clinicians with more than 5 biopsy proven patients with HSPN to join our project

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

3B. Despina Eleftheriou: Update of the KD-CAAP study "Multi-centre, randomised, open-label, blinded endpoint assessed, trial of corticosteroids plus intravenous immunoglobulin (IVIG) and aspirin, versus IVIG and aspirin for prevention of coronary artery aneurysms in Kawasaki disease."

KD-CAAP trial started opening up centres and will soon roll out recruitment across Europe. Trial run by MRC CTU at UCL and supported by C4C network

For further information please contact: PI - Despina Eleftheriou (d.eleftheriou@ucl.ac.uk), and PI - Paul Brogan (p.brogan@ucl.ac.uk)

3C. Despina Eleftheriou, Paul Brogan "Update on MYPAN study"

MYPAN results presented that support MMF as an alternative therapy to cyclophosphamide for childhood PAN. Paper in preparation.

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

3D. Seza Ozen: Comparison of pediatric criteria with adult classification criteria in granulomatosis with polyangiitis

- Multicentre study, at least 2 patients with GPA per center, and 2 controls (patients with other primary systemic vasculitis or mimicks such as sarcoidosis) per 1 GPA patient.

This project will start once we have the approval to sue the adult criteria.

For further information please contact: PI – Seza Ozen (sezaozen@gmail.com)

PRoS/PRINTO research grant

2020: deadline for submission December, 2020. Detailed info will be available on PRoS and PRINTO websites

Funds will consist of 10,000/year (total 30,000) in cash provided by PRoS, and of an equivalent amount in manpower for implementation and infrastructure provided by PRINTO.

Each PRoS/PRINTO grant will be funded for a maximum of 3 years. Proposals will be peer reviewed by members of the PRoS Council and members of the Advisory Council of PRINTO.

Sincerely yours

Marija Jelusic, Chair of Vasculitis WP