

Paediatric Vasculitis Working Party

Chair: Marija Jelusic

Secretary: Teresa Giani

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 19th, from 09:00 to 10:00, during PRs 2021 e-Congress.

1. CLINICAL CARE ACTIVITIES

George Reynolds –“The progress of the pedEUVAS registry”

in the EUVAS registry, the development of the pedEUVAS register has begun. 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.

Status of EUVAS/Vifor registry: 7 countries are planning to use a registry and consist of same team as involved in FAIRVASC project

Progress of the pedEUVAS registry since last year: PVAS has not started yet and requires modification to BVAS while CHAQ 1st draft released for clinical review

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

2. SCIENCE AND RESEARCH ACTIVITIES

2A. Mario Sestan, Marija Jelusic: „Treatment of patients with severe cutaneous manifestations in IgA vasculitis – multicenter, international study,,

Aims of the study: to evaluate the prevalence of severe skin manifestations (bullae, ulcerations, necrotic lesions) in children with IgA vasculitis, to propose a treatment for this group of patients since there is no enough data in the literature, to describe the clinical course, laboratory features and outcome of these patients

Patients and methods: observational retrospective multicenter none-profit study, pediatric patients diagnosed with IgAV according to EULAR/PRINTO/PRES criteria who developed severe cutaneous manifestations (bullae, ulcerations, necrotic lesions), disease onset before 18 years of age

Administrative aspects: study started in July 2021, data will be collected retrospectively until February 2022; 9 centers have sent the data and included in research (Turkey, Italy, Finland, Croatia).

The study is open for multicenter, international collaboration.

For more information please contact_Pls: Marija Jelusic (marija.jelusic@mef.hr) and Mario Sestan (mario.sestan@gmail.com)

2B. Nuray Aktay Ayaz, Figen Cakmak: "The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease"

Aims of the study: to evaluate the microvascular involvement in juvenile Behçet's disease, to find out the correlation between clinical findings and microvascular involvement, to find out the correlation between Behçet's disease activity scores and microvascular involvement, to compare nailfold capillaroscopic alterations and nailfold videocapillaroscopy scores of patients diagnosed with Behçet's disease with healthy volunteers

Patients and methods: patients aged 5-18 diagnosed with Behçet's disease according to the following criteria: Pediatric Behçet's disease criteria, International study group (ISG) criteria, The international criteria for Behçet's disease, - activity of Behçet's disease measured by Behçet's disease current activity form (BDCAF); laboratory findings: CBC, ESR, CRP, patergy testing, HLA B51 typing

The study is open for multicenter, international collaboration.

For more information please contact Pls: Nuray Aktay Ayaz (nurayaktay@gmail.com) and/or Figen Çakmak (figenatamancakmak@gmail.com)

2C. Despina Eleftheriou, Paul Brogan: 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 progress: 6 patients recruited, all NCA and EC submissions made, at least 20 sites to open

Target: 262 patients!

The study is open for multicenter collaboration.

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

2D. Seza Ozen, Ezgi Deniz Batu: Comparison of Ankara 2008 and adult classification criteria in patients with granulomatous polyangiitis (GPA)

- 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 use the adult criteria

For further information please contact Pls – Seza Ozen (sezaozen@gmail.com), Ezgi Deniz Batu (ezgidenizbatu@yahoo.com)

2E. Maria Vincenza Mastrolia: Comparison and performance of Kobayashi and Kawanet IVIg resistance scores in Kawasaki disease (the KIWI study)

- a retrospective-prospective, observational, international and multicenter study

Aims of the study: - to analyze demographic, clinical, biological and echocardiographic variables associated with IVIg resistance in European and North Indian cohorts of Kawasaki disease in children, to assess difference in clinical and treatment outcomes between European and North Indian Kawasaki disease cohorts, to assess the performance of Kobayashi and Kawanet scores in European and North Indian Kawasaki disease cohorts

The study is open for multicenter, international collaboration. A PRINTo feasibility survey will be forwarded to all PReS members

For further information please contact Pls: Maria Vincenza Mastrolia (maria.mastrolia@unifi.it) and/or Vignesh Pandiarajan (vigimmc@gmail.com)

3. EDUCATIONAL AND TRAINING ACTIVITIES

Teresa Giani announced the 1st PReS Educational Course "Vasculitis in paediatric age" a 1,5 day hybrid meeting in the February 2022, comprehensive training on main systemic vasculitides, new multidisciplinary diagnostic approaches, latest scientific advances, recent recommendations and guidelines for the

treatment. International expert speakers. The registration will be online (PReS website), limited subscriptions available. More information will be soon available on the PReS website!

Write us your interest/questions: *Teresa Giani* (teresa.giani@gmail.com), *Marija Jelusic* (marija.jelusic.drazic@gmail.com)

Rolando Cimaz—reported **the First European Congress on Kawasaki Disease EURO – KiDs Congress, 14 to 16 January 2021, Paris, France**. It was 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. There was 240 registered participants, more than 80 abstracts and 33 nationalities with participants from 5 continents and 31 talks.

4. ELECTION OF THE CHAIR AND SECRETARY

The new Secretary was elected: Teresa Giani, Anna Meyer Children’s Hospital, Florence, University of Milan, Milan, Italy

The Chair was re-elected: Marija Jelusic, University of Zagreb School of Medicine, University Hospital Centre Zagreb, Zagreb, Croatia

Sincerely yours,

Marija Jelusic, Chair of Vasculitis WP

Teresa Giani, Secretary of the Vasculitis WP