pediatric rheumatology european society

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

October 9, 2022

To all Vasculitis Working Party members

Dear Colleagues,

Behind us is an active and project-rich year in which PReS Vasculitis Working Party members worked hard and thus we finished 3 projects, there are updates of 4 ongoing projects and we also have the announcement of some new ones.

So, please kindly find a short report of our Vasculitis Working Party meeting that we had on September 20th, from 15:15 to 16:15, during PReS 2022 Congress as a hybrid event.

1. ELECTIONS WITHIN THE WP

The Core team of the Vasculitis WP :
1. Chair: Marija Jelusic (Zagreb)
2. Secretary: Teresa Giani (Florence)
3. Lead of education and training: Teresa Giani (Florence)
4. Lead of clinical care: Neil Martin (Glasgow)
5. Lead of science and research: Ezgi Deniz Batu (Ankara)
- EMERGE representative: Mario Sestan (Zagreb)

- Currently, there were changing from 3 years to 4 years tenure position which align this with PRES Council positions
- Traditionally the voting process takes place live at the PReS congress during the WP meeting, beforehand WP members are asked via email to candidate themselves if interested. In 2023¸. during the PReS congress we will have election of the Core team. Please feel free to candidate yourself. Our mailing list contains 145 Vasculitis WP members (including 6 new members from this year) from all around the world

2. EDUCATIONAL AND TRAINING ACTIVITIES

Teresa Giani, Jordi Anton, Marija Jelusic: Summary of the **1st PReS Vasculitis Course - In memoriam Rolando Cimaz, hybrid, 28-29 March 2022, Barcelona** Sant Joan de Déu Barcelona Hospital)

-An exhaustive overview of clinical presentations of major primary vasculitides was initially offered, in addition to specific laboratory, histopathological and radiological data to refine skills for diagnosis. COVID 19-related vasculitis, primary CNS vasculitis, and newly identified monogenic disorders associated with systemic vasculitis have been discussed. Monitoring of disease activity and damage, recent updates on management guidelines, best clinical practices, and new pharmacological options have been detailed on the last day of the course together with a lecture on how to critically read an article on pediatric vasculitis.

- Eight interactive clinical cases have been presented by young doctors pointing out the potential role of pitfalls and mimickers in the diagnostic process. Two best clinical case presenters (Dr. Nolvia Castillo, Dr. Mario Sestan) have been selected and awarded with the free registration for the PReS 2022 congress
- 134 participants from all around the world attended the course, 30 participants were in person, 17 invited speakers

CliPS Project: collection of real life clinical practice strategies from worldwide physicians through the questionnaire of the following medical conditions: lupus nephritis, Kawasaki disease, IgA vasculitis, monogenic autoinflammatory diseases, PFAPA, SURF, sJIA/Still's disease
contact: info@jircohorte.ch, www.jircohorte.org/clips

3. SCIENCE AND RESEARCH ACIVITIES

FINISHED PROJECTS (2021/2022) - MANUSCRIPTS UNDER REVISION

1. Marija Jelusic, Nastasia Kifer *"Histological predictors of outcome in patients with Henoch-Schonlein purpura / IgA vasculitis and nephritis"*
- collaboration between WP members from Croatia, Italy and Israel
- included 67 renal biopsies from 2003 until 2021
- for the first time compared the four most frequently used classifications for IgAV nephritis
- SQC proved to be the best, followed by Oxford classification; cellular crescents in SQC, mesangial hypercellularity and tubular atrophy in Oxford have shown significant contributions in outcome prediction
Kifer N, Bulimbasic S, Sestan M, Held M, Kifer D, Srsen S, Gudelj Gracanin A, Heshin-Bekenstein M, Giani T, Cimaz R†, Gagro A, Frkovic M, Coric M, Jelusic M. „SQC and Oxford classifications predict poor renal outcome better than ISKDC and Haas in patients with IgAV nephritis: a multicenter study" - manuscript under revision

2. Ezgi Deniz Batu, Seza Ozen *"COVID-19 associated pediatric vasculitis study"*
- included 42 patients from 14 pediatric rheumatology centers
- the most common clinical manifestation of COVID-19 associated vasculitis was skin involvement and the most frequent diagnosis was COVID-19 associated IgAV
- COVID-19 associated IgAV had more severe disease course than non-COVID-19 associated IgAV: more prevalent renal involvement and less prevalent complete recovery.
Ezgi Deniz Batu, Seher Sener, Gulcan Ozomay Baykal, Elif Arslanoglu Aydin, Semanur Özdel, Alenka Gagro, Fatma Gül Demirkan, Esra Esen, Nilufer Akpınar Tekgöz, Kubra Ozturk, Olga Vougiouka, H. Emine Sonmez, Merav Heshin-Bekenstein, Maria Cristina Maggio, Ummusen Kaya Akca, Marija Jelusic, Aysenur Pac Kısaarslan, Banu Çelikel Acar, Nuray Aktay Ayaz, Betül Sözeri, Seza Özen. „The characteristics of patients with COVID-19-associated pediatric vasculitis: An international, multicenter study" - manuscript under revision

3. Mario Sestan, Marija Jelusic *"Clinical features, treatment and outcome of patients with severe cutaneous manifestations in IgA vasculitis - multicenter study"*
- included 64 patients from 13 paediatric rheumatology centers with the most severe skin changes in IgAV
- IgAV patients with severe skin manifestations more frequently developed IgAV nephritis, had worse outcome of renal disease and were more frequently treated with systemic glucocorticoids and were more likely to develop the most severe gastrointestinal manifestations
Sestan M, Kifer N, Sozeri B, Demir F, Ulu K, Silva Clovis, Campos Reinan T, Batu ED, Koker O, Sapina M, Srsen S, Held M, Gagro A, Fonseca A, Rodrigues M, Rigante D, Filocamo G, Baldo F, Heshin-Bekenstein M, Giani T, Kataja J, Frkovic M, Ruperto N, Özen S, Jelusic M „Clinical features, treatment and outcome of patients with severe cutaneous manifestations in IgA vasculitis: multicenter international study"-manuscript under revision

ONGOING RESEARCH PROJECTS - UPDATE

A. Ummusen Kaya, Seza Ozen: 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 started in December 2021
- objectives of the study: comparison of newly identified adult classification criteria to pediatric classification criteria in patients with granulomatous polyangiitis, to validate the newly identified adult classification criteria in children, to evaluate the sensitivity and specificity of adult classification criteria in pediatric patients

If you are interested to participate, for further information please contact PIs – Seza Ozen (sezaozen@gmail.com), and Ummusen Kaya (ummusenkaya@gmail.com)

B. 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: , all NCA and EC submissions made, at least 20 sites to open

Target: 262 patients!

The study is open for multicenter collaboration.

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

C. Nuray Aktay Ayaz, Figen Çakmak: "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

-Project progress: the enrollment of the study is extended since the ethical comittee approval was obtained on April 2022, till now included 37 patients from 6 Turkish centers, preliminary results: neoangiogenesis was found to be significantly more common in the NVC evaluation of patients with lower hemoglobin values at the time of diagnosis (p=0.014)

- The study is open for multicenter, international collaboration

For more information please contac PIs: Nuray Aktay Ayaz (nurayaktay@gmail.com) and Figen Çakmak (figenatamancakmak@gmail.com)

D. Maria Vincenza Mastrolia, Vignesh Pandiarajan: 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
- project progress: 112 enrolled patients, 4/5 participating core members obtained EC approvals
- an official reminder to speed up EC submission will be sent out to the satellite sites the next month
- next steps and deadlines:

October 2022: Patient recruitment and data entry of the 5 core members, December 2022: Ethics committee approval of further satellite centers, Jnuary 2023: Patient recruitment and data entry of satellite members, April-May 2023: KIWI study participants web-meeting, June 2023: Half patient sample reached, total sample size 246
The study is open for multicenter, international collaboration.
For further information please contact: Maria Vincenza Mastrolia (maria.mastrolia@unifi.it) and Vignesh Pandiarajan (vigimmc@gmail.com)

4. NEW RESEARCH PROJECTS (PROPOSALS)

1. Fatma Gül Demirkan, Nuray Aktay Ayaz: *"Biologic Drug Strategies in Pediatric-Onset Behçet's Disease"*

- aims of the study: to evaluate the characteristics of biologic treatment regimens, their efficacy on any clinical involvement of BD, their side effects and outcomes
- inclusion criteria: patients < 18 years, follow-up period of >1 year, at least one biologic drug for at least 3 months, satisfying at least one of the following international BD criteria: International team for the revision of the International criteria (ICBD)/pediatric Behçet's disease (PEDBD) study group

- to create a comprehensive overview of the use of biologic drugs in pediatric-onset BD on which does not have a consensus globally
- at least 2 patients for each center

The study is open for multicenter, international collaboration.
For further information please contact: Özlem Akgün (drozlemakgun@hotmail.com), Fatma Gül Demirkan (fatmagy@gmail.com) and Nuray Aktay Ayaz (nurayaktay@gmail.com)

2. David Cabral: Study for potential collaboration CARRA - PReS (or individual PReS members) ***„To comparatively evaluate CARRA-endorsed Consensus Treatment Plan (CTP) alternatives for pAAV (GPA/MPA) in the PedVas registry"***

- how to participate:
  - join PedVas (IRB +/- contract) - contact Else at ebosman@cw.bc.ca; dcabral@cw.bc.ca
  - commit to using specified regimens for remission-induction and remission-maintenance treatment of moderate-severe GPA/MPA:
  - limited dosing and duration of IV and oral prednisone
  - train in use of PVAS (with testing) if not proficient using on-line training module
  - go to: https://redcap.link/PVAStraining to leave your information to create an account
  - capture damage with pVDI PLUS additional specific drug toxicities
  - collect clinical data +/- biological samples at specified times according to schedule

For further information please contact PI: David Cabral (dcabral@cw.bc.ca

3. Marija Jelusic *„Consensus conference on the definition of disease status, outcomes and follow-up in IgA vasculitis"*
- observational retrospective multicenter none-profit study
- aims of the research:

- to define the clinical course of IgA vasculitis with the aim of reaching a consensus in the definition of the outcome of IgA vasculitis and IgA vasculitis nephritis
- to define disease status in IgA vasculitis categorized as complete remission, partial remission, progressive disease or relapsing disease
- to define persistence of nephritis and purpuric rash

- to define follow-up for patients with higher risk for renal involvement, and for patients with nephritis and low, moderate and high risk for progression of nephritis

- inclusion criteria: pediatric patients with diagnosis of IgAV, according to the Ankara 2008 EULAR/PRINTO/PReS criteria

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

Dear PReS Vasculitis Woring party members, please feel free to join in some ongoing and new research projects.

Sincerely yours,

Marija Jelusic, Chair of the Vasculitis WP